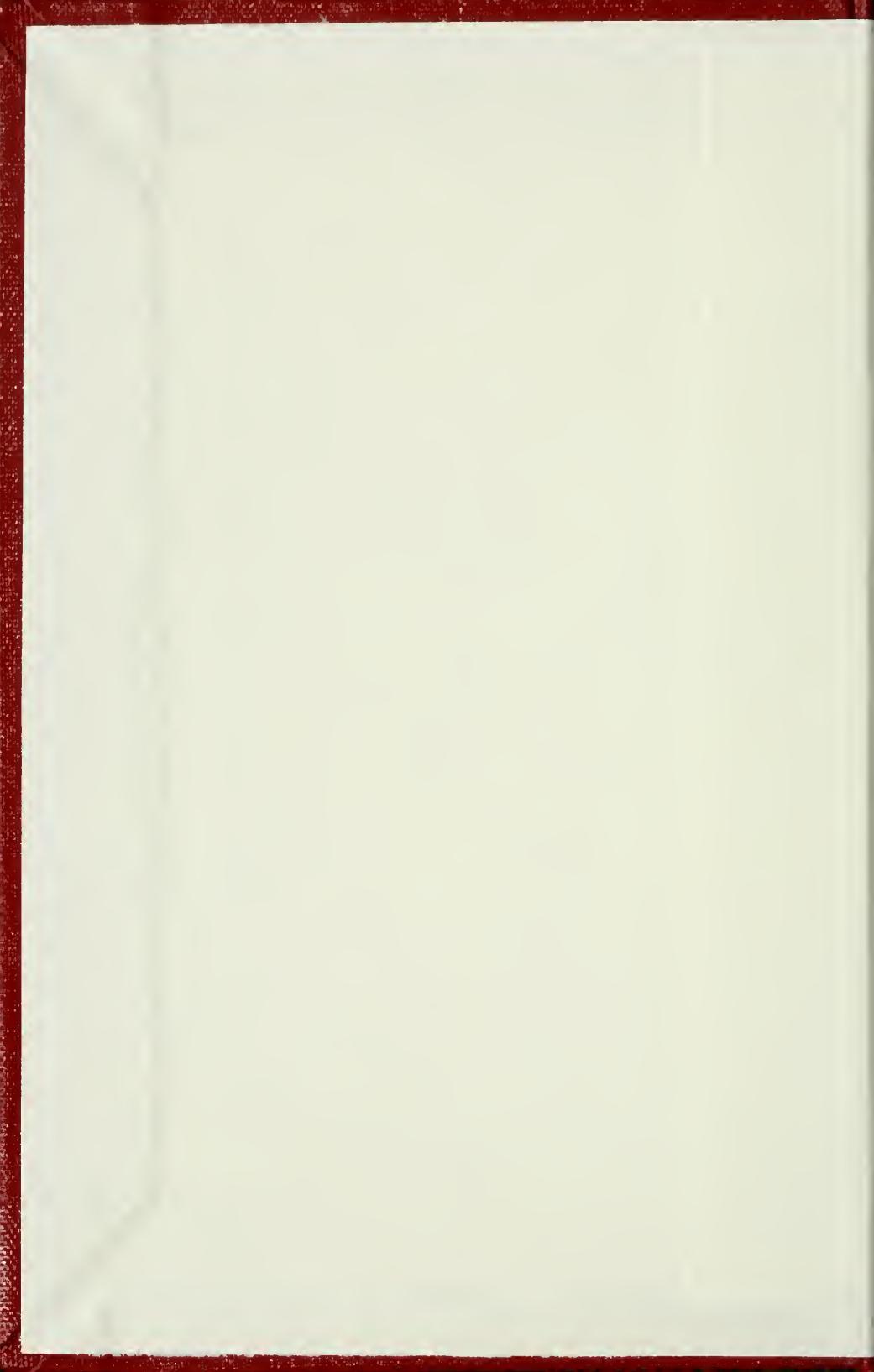




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THE DISEASES AND DEFORMITIES OF THE FŒTUS:

AN ATTEMPT TOWARDS

A SYSTEM OF ANTE-NATAL PATHOLOGY.

BY

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WITH PLATES.

VOLUME II.

CONGENITAL DISEASES OF THE SUBCUTANEOUS TISSUE AND SKIN.

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TO

ALEXANDER RUSSELL SIMPSON,

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PROFESSOR OF MIDWIFERY AND DISEASES OF WOMEN AND CHILDREN
IN THE UNIVERSITY OF EDINBURGH

1870—1895,

THIS VOLUME IS SPECIALLY DEDICATED

BY HIS FORMER ASSISTANT,

THE AUTHOR.

P R E F A C E.

IT is now two and a half years since the first instalment of my ATTEMPT TOWARDS A SYSTEM OF ANTE-NATAL PATHOLOGY came from the press; and this, the second volume, has had its appearance delayed by causes which have been unavoidable. Chief amongst these retarding causes has been the unexpected magnitude of the task of gathering together and classifying the numerous contributions which have been made to foetal pathology during the last fifty years; for it is now more than half a century since Graetzer's book was published, and it must still be regarded as the most comprehensive and authoritative completed work on the diseases of the foetus. I have, therefore, had to face the question whether, in view of the vast amount of accumulated information relating to ante-natal disease, I should endeavour to compress the consideration of all the remaining maladies of the foetus into the three volumes originally set aside for this purpose in the preface to the first part, or whether I should continue to treat all the subjects in the same exhaustive manner as I did general foetal dropsy, and so depart from the limits of the work announced two years ago. I have, after considerable thought, chosen the latter alternative, even although doing so has led me to face the possibility that I may not be able to complete my task within the limits of time which I may reasonably expect to have at my disposal; for I have felt that it is better to discuss fully a part even of the subject, than to consider in an imperfect and inadequate way

the whole. I might, of course, have considerably lessened the task by omitting the neonatal maladies ; but these, I have always felt, are so closely associated with foetal diseases proper as to be practically inseparable, and certainly the study of the one group is sure to aid materially the understanding of the other.

In the present volume, therefore, and for the reasons above mentioned, I have proceeded with the consideration of the congenital diseases of the subcutaneous tissue and skin. A reference to the last Chapter (XIII.) will enable my readers to estimate how much of this part of the subject I have been able to overtake, and how much still remains to be discussed before the ante-natal maladies of the osseous system can be begun.

There is a second and scarcely less important cause that has delayed the appearance of this volume. It is the fact that without any special training in dermatology, and with comparatively few opportunities of seeing cutaneous maladies, I have had to undertake the description and classification of the congenital diseases of the skin, affections which confessedly present many perplexing problems for solution. It was, therefore, necessary for me to acquaint myself with the most recent and authoritative writings on cutaneous diseases, and this occupied time. Even as it is, I must ask the forbearance of the specialists in dermatology who may read this work and find in it errors such as could not have been committed by one specially versed in dermatological literature and practice. Whilst these sheets have been passing through the press, Unna's monumental work on the "*Histopathologie der Hautkrankheiten*" has appeared, and therein will be found a special section dealing with certain of the congenital skin diseases (Missbildungen). Had Unna's book been published earlier, I should probably have chosen to adopt that master's plan of arrangement in preference to the

one which I have constructed, and which will be found indicated in the concluding chapter of this volume. At the same time, it must be remembered that whilst I have attempted to gather together all the diseases of the skin to which the term *congenital* could be correctly, if only occasionally applied, Unna has separated for special consideration only such as may properly be designated cutaneous deformities or malformations.

This volume I have ventured specially to dedicate to Professor A. R. Simpson, who this year celebrates the twenty-fifth anniversary of his appointment to the Chair of Midwifery and Diseases of Women and Children in the University of Edinburgh. I may here again acknowledge the constant kindness and encouragement that I have received at his hands, both when I had the good fortune to serve as his assistant, and since.

In conclusion, it may be stated that with the exception of the drawings illustrating sclerema and œdema neonatorum, which are from specimens in my own possession, the Plates in this volume are copied from the works of Kyber, Caspary, Straube, and Arndt.

J. W. BALLANTYNE.

24 MELVILLE STREET, EDINBURGH,

April 1895.

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THE DISEASES OF THE FŒTUS.

CHAPTER I.

SCLEREMA NEONATORUM.

IDIOPATHIC DISEASES, *continued from Vol. I.*

DISEASES OF THE SUBCUTANEOUS TISSUE, *continued*—SCLEREMA NEONATORUM: DEFINITION; SYNONYMS; HISTORICAL SKETCH; VARIETIES; DESCRIPTION OF CASES E AND F.

A STUDY of the conditions known as sclerema neonatorum and œdema neonatorum affords a most convincing proof—if such, indeed, be wanted—of the unsatisfactory nature of our knowledge of the maladies of the neonatal state. A review of the literature of the subject forces us to ask whether authors have been describing one disease or many under the name of sclerema, and whether there is any reasonable hope of introducing order into the confused mass of facts, clinical and pathological, that has been handed down to us by previous writers. Truly, if the literature were less extensive the task would be easier, for it is now next to impossible to eliminate observations which had reference to conditions having little in common with sclerema.

It is particularly difficult for a British author to obtain a correct conception of the subject, for the disease is exceedingly rarely met with in this country; and although I have been fortunate in seeing one instance of it (the only one, indeed, that has occurred in the Edinburgh Maternity Hospital for many years), yet my opportunities of study have been almost insignificant when compared with those of some Italian observers, who have been able to carry out as many as seventy autopsies on scleremic infants annually. For the same reason I hesitate to press my own views prominently forward, especially since they are in

some respects at variance with those of authors who have had immensely greater facilities for the study of this morbid process.

In order to remove, as far as possible, some of the confusion which has surrounded the whole subject, it may at once be stated that sclerema neonatorum has nothing in common with the skin disease known as scleroderma. This conclusion is warranted, for there is now a consensus of opinion, which, to my mind, embodies the truth, to this effect. Further, the disease is essentially different from erysipelas neonatorum, and it is not, I think, to be confounded with the pathological state which so often occurs in atrophic infants perishing from cholera infantum or summer diarrhoea. Further, whilst I am convinced that there is one morbid condition to which the name "sclerema" ought preferably to be applied, and another which should be designated "œdema neonatorum," I am ready at the same time to admit that the differentiation will be a matter of some difficulty, especially if it be conceded (and the observations of G. Somma render the concession necessary) that sclerema is not always characterised by induration of the subcutaneous tissue, but sometimes by oedematous infiltration of the same.

Some further remarks upon the relation of sclerema to œdema neonatorum will be made immediately, when I come to consider the definition which has been formulated for the former condition.

Sclerema neonatorum is a condition which, whilst only occasionally present at birth, occurs always so soon thereafter, and so often in premature infants, that its omission from this work would be unpardonable.

Definition.—From what has been already said, it will have been gathered that it is impossible, in the present state of our knowledge, to frame a satisfactory definition of sclerema neonatorum. In this respect it is neither in a better nor in a worse condition than many other foetal and neonatal maladies. The following definition must, therefore, be regarded as purely provisional, and it is quite probable that the results of future investigations may very considerably modify it.

Sclerema may be described as *a grave disease, occurring almost*

always in the new-born infant, rarely found in this country, but more often met with in France and Italy, characterised by induration, and sometimes by œdema, of the subcutaneous cellular tissue and by lowering of the body-temperature, and due possibly to some trophic lesion of the nervous system. œdema neonatorum, on the other hand, may be defined as a symptomatic condition, characterised always by serous infiltration of the subcutaneous tissue, probably caused in most cases by cardiac, vascular, renal, or pulmonary disease in the new-born infant. Sclerema I regard, therefore, as a disease *per se*, whilst I look upon œdema neonatorum as a symptom of several morbid states rather than as a distinct pathological entity. Sclerema is a malady practically peculiar to the congenital and neonatal state; œdema neonatorum is simply œdema occurring in the new-born, and caused by the same factors as produce it in adult life. When I wrote in 1889, I was of opinion that sclerema would always be found characterised by induration of the subcutaneous tissue, and that in the cases in which œdema was present adequate causes for the same, identical with those producing œdema in the adult, would be discovered.* G. Somma (150),† a recent writer, with wide experience of sclerema, has, however, pointed out that cases characterised by œdema instead of induration of the subcutaneous tissue, but otherwise having all the features of sclerema, occasionally occur, and that in them no such cause for the œdema as I had pointed out existed. "Sclerema," he says, "is a neurosis of the thermic centres, occurring always in the new-born, on account of which, along with the rapid and progressive lowering of the body-temperature, there may follow either œdematosus infiltration of the subcutaneous connective cellulo-adipose tissue, or the induration of the same, or both the said forms occurring at the same time." Somma, in his definition of this puzzling malady ("enigmatica infermita"), regards rapid and progressive lowering of the body-temperature

* In my paper (*v. Brit. Med. Journal*, p. 404, Feb. 22, 1890), sclerema was "provisionally defined as a rare disease, occurring most commonly in the new-born infant, characterised by induration of the subcutaneous tissue, and being little amenable to treatment." It was added that "the primary pathological factor may be, and probably is, a trophic lesion of the nervous system."

† The figures within parentheses refer to bibliography at the end of Chapter IV.

and not any change in the subcutaneous tissue, as *the distinctive character of sclerema*. In my first definition of sclerema I looked for a distinguishing feature in the state of the subcutaneous tissue, and not in that of the temperature of the infant. The consideration of Somma's excellent monograph, with its wealth of clinical material, has caused me to modify my first opinion of sclerema, without, however, leading me to change my views as to what cases should be included under the designation "œdema neonatorum." Whilst I am willing to regard the information gained by the use of the thermometer as of first importance in the recognition of *sclerema*, and whilst the clinical evidence brought forward by Somma and others has led me to admit the existence of an œdematos variety of this malady, I am still of opinion that the term *œdema neonatorum* should be definitely reserved for the cases in which anasarca occurs as a symptom of cardiac, vascular, renal, and pulmonary disease in the new-born infant. The use of *œdema neonatorum* as a synonym for sclerema—a procedure which has been very common with authors, as will be immediately shown—has introduced into the whole subject a great deal of confusion, and the abandonment of its use in such a sense cannot but be beneficial.

Synonyms.—Few diseases have had the good (or rather bad) fortune to have had so many different names given to them as has sclerema. This has been due partly to the differences which exist in its clinical manifestations and partly to the resemblances which it bears to some other maladies.

The first specimen was graphically named "foetus vivus, frigidus et rigidus" (1), and Underwood (219) called the malady an "abnormal erysipelatous inflammation," although later he adopted the expression "skin-bound" or "hide-bound disease." Andry (5) was the first to give to it the designation "endurcissement du tissu cellulaire des enfans nouveaux-nés," a phrase which is still frequently used, especially in Germany, where it appears under the form of "Zellgewebsverhärtung Neugeborenen." The English equivalent is "induration of the cellular tissue;" the Italian, "indurimento del tessuto cellulare;" and

the Latin, "induratio" or "stipatio telæ cellulosa," or "induratio textus cellulosi neonatorum." Another group of names had its origin in the term "sclérôme," proposed by Chaussier (169). Variations of this were "sclérome," "sclérémie," and "sclerisma," in French; "sklerem," "acute sclerose," "sklerysma," and "Haut-sklerom," in German; "sclerema," "sclerosi," "scleromia," "scleroma," and "scleremia," in Italian; and "esclerema" and "esclerosis" in Spanish. English writers have generally adopted the term "sclerema neonatorum," and it may, in fact, be regarded as the favourite expression in all countries except in Germany, where it is condemned as unclassical.

"Œdema neonatorum," with its variations, has been also employed by writers, some of whom have regarded it as a synonym of sclerema, whilst others have considered the condition so named as pathologically distinct from sclerema, whilst yet others have used it to express a variety of that disease. Those who employed it as a synonym usually added a qualifying adjective; thus Léger (37) in 1825 used the expression "l'œdème compacte" ("edema compatto," Ital.); and other names have been "œdématie concrète" ("edemazia concreta," Ital.), and "edema algido." Valleix (65), in 1835, proposed the descriptive phrase "l'asphyxie lente chez les enfans nouveau-nés," and L. Somma (113) suggested that of "algidismo essenziale dei bambini" in 1875. "L'algidité progressive chez les nouveau-nés" had been used by Hervieux (87) in 1855.

"Scleroderma neonatorum" has occasionally been employed, but it is a misleading name. "Squirrosarque" (Fr.); "gespannte Haut" and "Greisenhaftigkeit" (Germ.); "serrone" (Ital.); and "cutis rigor" (Latin), are still other terms that have been applied to this disease. "Sclerödema," used by Totenhofer (127) and Soltmann (215) is the most recent addition to the already long list of synonyms.

Historical Sketch.—Sclerema neonatorum has no written history prior to the beginning of the eighteenth century. This fact is surprising, for it is impossible to doubt its occurrence in earlier times; but the absence of an early sclerema literature may be explained, in part at least, by the want of interest in the diseases

of children, and more especially of new-born infants, which then prevailed. It is nevertheless somewhat strange that no hint of the existence of this disease should be found in the classic writings of Hippocrates, Galen, Aretaeus, Paulus Aegineta, and others.

The history of the subject may be divided into two periods, the first corresponding to the eighteenth, and the second to the nineteenth century. At this point I shall indicate only the chief contributions that were made to the growth of our knowledge regarding sclerema.

1. *In the Eighteenth Century.*—The history of sclerema in the last century began with the record of the classical case of Usenbenzius (1) in 1718. It was of special interest, for the disease was noted by the midwife at the time of birth, and was, therefore, truly foetal. Usenbenzius was much struck by the marble-like coldness of the skin and the rigidity of the limbs; and in keeping with the beliefs of the time he found a possible cause in a maternal impression. The infant, a female, was prematurely born (eighth month) and died in six hours. No further contribution was made to the study of this disease for many years, but Schurig (212), in 1732, and a few others made reference to Usenbenzius's case. Underwood (219) was the next author of any note who wrote upon this disease; but he seems at first to have confused it with erysipelas. Nevertheless his description of its clinical features was a good one, and in the later editions of his treatise on the Diseases of Children he clearly differentiated the malady from erysipelas, and also pointed out the distinguishing features between the condition described by him and that seen in Paris by Andry. Doublet (4), writing in 1785, found that in the instances met with by him the infants were syphilitic, and concluded somewhat hastily that it was a symptom of syphilis. Andry (5), having seen certain cases of serous infiltration of the subcutaneous tissue, and having read Underwood's work, came to the conclusion that the two conditions were the same, and so introduced some confusion into the subject; but his contribution was a notable one, and did good service by pointing out that the disease had nothing to do with

syphilis. Andry's communication, and that also of Auvity, (6), were specially of value for the rich store of clinical details that they contained. Auvity's memoir may, in the opinion of G. Somma (150), be read even at the present day with "great interest and immense profit." Souville (7) wrote on the influence of the surroundings of Calais on the development of the disease there, and Hulme (8) pointed out the frequency of pulmonary complications. Moscati (9) in Italy, and Girtanner (186) in Germany, also described the malady.

The contributions of the eighteenth century were not numerous, and were almost purely clinical; in a few instances the results of autopsies were given, but it was for their symptomatological details that they were of most value. A perusal of them shows that even at this early date the two varieties, the indurative and the cedematous, were under observation, and that on this account great confusion already existed in the minds of observers.

2. *In the Nineteenth Century.*—During the present century many separate articles have appeared treating of sclerema, and most of the well-known text-books upon pediatrics and skin diseases have given some space to its consideration. I shall here do little more than indicate some of the outstanding features of this extensive literature, and refer the reader to the end of Chapter IV. for the full bibliography of the subject.

Amongst the contributions of the first decade of this century may be mentioned those of Frank (11), Horn (16), Stütz (14), and Lodemann (17). Frank, in his *Reise nach Paris*, gave an interesting account of sclerema as met with in the Parisian Hospitals, and suggested that many of the cases described were really instances of erysipelas and tetanus. The seventeen cases also which were reported by Horn in Germany have been considered by Hennig (190) and others as examples of erysipelas neonatorum and not of sclerema. Lodemann attempted to differentiate two forms of the disease according as the skin and cellular tissue or the muscles were affected with induration, and pointed out its resemblance to phlegmatia alba dolens of the puerperium.

During the second decade of the century Liberali's observa-

tions (29) occupied a prominent place, both for their extent and value: the post-mortem appearances were fully described (as many as seventy autopsies were made in one year), and paralysis of the subcutaneous nerves by cold was advanced to explain its mode of origin. Loder (21) noted its frequency in the cold months of the year; Feiler (182) said it was most often congenital, and thought that it had some connexion with syphilis; Troccon (22 and 24) and Dugès (176A), like Hulme, advanced pneumonia as a causal factor; Carus (25) compared it to marasmus senilis and the condition popularly known as "dead fingers;" Jahn (194) thought it was the same as *morbus cœruleus*; and Kutsch (26) looked upon it as a primary disease of the lymphatic system. Sybel's case (19) was probably one of erysipelas. Heriveux (87) seems to have seen a congenital example of sclerema.

Between the years 1820 and 1830 appeared, amongst others, the contributions of Léger (37), Henke (189), Carminati (35), Palletta (38), Denis (40), Siebold (42), Heyfelder (192), Marzari (41), and Billard (53 and 163). The views brought forward by these writers differed to an extraordinary degree, and it may be said that confusion reigned paramount. Carminati regarded sclerema as an inflammatory morbid process; Palletta thought it was due to want of expansion of the lungs; and Denis found that it frequently occurred in association with gastro-intestinal disturbance, and believed it to be an inflammatory affection of the subcutaneous tissue and of the intestine, an entero-cellular phlogosis. Léger concluded from some experiments of Chevreul* that the subcutaneous tissues of the new-born infant were easily coagulable by cold, and founded upon this an etiological theory; but he also regarded shortness of the intestine (a condition which he found in his autopsies) as a causal factor. Zimmermann (39) thought that the sclerema of the French authors was erysipelas, and that it had no connexion with the "skin-bound disease" of the English writers. Billard separated sclerema from œdema, and regarded the former as a cadaveric phenomenon, or at least one occurring in *articulo mortis*, and the latter as a simple œdema analogous to that met with in adults, and produced by certain predisposing and exciting causes (congenital debility,

* Chevreul, *Considérations génér. sur l'analyse organique*, p. 218, Paris, 1824.

plethora, obstructed circulation, etc.). His views dominated in a marked degree those of the writers who followed him. Heyfelder (192) was of opinion that sclerema was due to a disordered state of respiration with imperfect oxygenation of the blood, and thought that it was closely allied to icterus neonatorum.

With the exception of the work by Valleix (65 and 221) no important communication was made to the study of sclerema between the years 1830 and 1840. The name given by Valleix to this disease (*viz.*, "l'asphyxie lente") indicated the idea he had formed of its nature; he believed it to be a general malady characterised by disturbances of the circulatory and respiratory functions, and caused by the action of cold. In many particulars his opinions and those of Ryan (211) resembled Billard's.

During the fifth decade of this century many writers of textbooks on pediatrics considered sclerema more or less fully: of these, Barthez and Rilliet (158), Barrier (157), and West (226) may be specially mentioned. Mildner (77) contributed an article in 1847, which was largely quoted by subsequent German writers; and Thirial (75) made a comparative study of scleroderma ("sclerema in the adult") and sclerema neonatorum.

Bouchut (164) recognised two varieties of sclerema, a simple and an oedematous: the former he regarded as comparable to scleroderma in the adult, and not to be confounded with adipose induration, and the latter was ascribed to a suspension of the capillary circulation in the skin. Elsässer (81), Clertan (82), Bednar (160), Legroux (84), Bierbaum (86, etc.), and Löschner (90) also contributed papers on sclerema between the years 1850 and 1860. Bednar dwelt upon the action of cold upon premature and weakly infants as the cause of oedema of the new-born; Legroux looked upon it as a syncopal asphyxia from muscular inertia; and Löschner, from the characteristic lowering of the body-temperature, called the disease "progressive *Algidität*."

Very little of any value concerning sclerema was written between the years 1860 and 1870. Isambert (100, 101, 103) contributed some cases, in one of which the patient was 13 months old; but it has been thought by some (*e.g.*, Crocker) that the disease in this instance was scleroderma occurring at an unusually early age.

The contributions of the eighth decade of the nineteenth century were noteworthy. They included articles by Vogel (223), Clementovsky (111), L. Somma (113), Cruse (114), Barlow (115), C. Hennig (97), Parrot (206), Steiner (217), and Depaul (122). Vogel looked for the primary etiological factor in defective innervation of the heart-muscle ; Steiner held somewhat the same opinion ; Marcacci (107) considered sclerema as the epiphomenon of a very complex infantile disorder ; and Hennig demonstrated the tendency of German thought upon this subject by affirming that induration of the cellular tissue was an acute inflammatory affection of the integument, and that the symptoms were due to venous stasis predisposed to by weakness of the infantile organism. Both Clementovsky and Depaul separated sclerema from oedema neonatorum and gave the symptoms of the differential diagnosis. The latter considered oedema to be due to stasis from congenital feebleness and other causes, whilst sclerema he regarded as a special variety of emaciation with almost complete disappearance of the fluid elements of the subcutaneous tissue. His views with regard to sclerema he got from Parrot, who stated that it was only one of the symptoms of athrepsia or infantile atrophy, whilst athrepsia he regarded as one of the complications of oedema neonatorum. Luigi Somma's contribution was one of great value, although it was strangely lost sight of by subsequent writers. He saw an enormous number of cases, and from their study drew the conclusion that sclerema and oedema were simply different forms of one morbid entity, and gave to them the names "sclerema duro" or "secco," and "sclerema edematoso" or "molle." The cause he sought for in a vasomotor and trophic neurosis produced by morbid stimulation of the sympathetic from cutaneous chilling due to atmospheric cold, etc., and consisting in an inertia of the bio-chemical processes of the body. My theory, which has some points in common with that of Somma, was advanced by me in 1889, without previous knowledge of the work of the distinguished Italian.

Since 1880 cases of sclerema have been reported by Soltmann (130 and 215), Namias (133), Porteous (140), Money (142), Barrs (143), Stephen Mackenzie (146), Northrup (148), and

others ; and articles dealing with the disease have appeared in the text-books of Henoch (191), Descroizilles (173), Ellis (179), D'Espine and Picot (175), Baginsky (156), Unger (220), Max Runge (2nd edit., 210), Ashby and Wright (2nd edit., 155), and others.

Langer (123), in 1881, pointed out that in new-born infants the panniculus adiposus, on account of the preponderance in it of palmitine and stearin, solidifies by the action of cold at a higher temperature than it does in the adult. This observation has been used by G. Somma (150) and others to account for the hardening of the tissues of collapsed scleremic infants. Henoch described sclerema and œdema neonatorum as separate diseases, and by sclerema he understood the indurative process occurring, as Parrot pointed out, in atrophic infants after continued diarrhoea. Soltmann differentiated between sklerödem and sklerema adiposum, and in this he has been followed by Max Runge; evidently the sklerema adiposum of Soltmann and Runge is Parrot's sclerema. G. Somma (150) does not consider this drying up of the tissues after an exhausting illness as true sclerema neonatorum, which he defines as a neurosis of the thermic centres, manifesting itself in three clinical forms,—an œdematosus, an indurative, and a mixed. Soltmann's sklerödem would correspond to G. Somma's sclerema. With regard to other contributions, it may be said, in conclusion, that Campbell (166) looked upon sclerema as probably a general tropho-neurosis, and as possibly allied to myxœdema in the adult ; Dumas (141) revived an old theory when he dwelt upon the probable identity of œdema neonatorum and phlegmatia alba dolens ; and Pavone (152) found in twenty-three cases microscopic appearances very similar to those described by me in 1889.

Whilst the contributions of the eighteenth century were chiefly of clinical importance, those of the nineteenth have served to form the basis of an understanding of its pathological anatomy and pathogenesis. A rich store of facts, gleaned from numerous autopsies, is now at our disposal ; and from this, and from the advances in our knowledge of the bio-chemical phenomena of neonatal physiology, it is reasonable to look for

a trustworthy explanation of the nature and mode of origin of sclerema neonatorum.

Varieties.—Even at an early period in the history of this subject, it began to be recognised that there were two closely allied conditions, to which the names sclerema, induration of the cellular tissue, and oedema neonatorum were being applied. Roughly speaking, the character of the one condition was induration of the subcutaneous cellular tissue, and that of the other oedematous infiltration. At first it was thought that these were different forms of the same disease—sclerema; and such names as “sclérème concret” and “sclérème oedemateux” were introduced. Soon, however, attempts to distinguish two separate diseases were made, and it was during this process that conditions foreign to sclerema, as we now understand it, were brought in to complicate matters. During the last few years the tendency has been to recognise an indurative and an oedematous variety of sclerema, and to get rid of the other morbid processes, such as cadaveric or preagonic hardening of the cellular tissue in wasting maladies, and oedema neonatorum due to causes similar to those causing oedema in the adult. G. Somma (150) recognises three varieties of sclerema, founded upon the state of the subcutaneous tissue: an *oedematous* (“forma edematosa”), an *indurative* (“forma dura”), and a *mixed* form (“forma mista”). He also enumerates three varieties, according to the extent of the morbid process: a *partial* or *circumscribed*, a *diffuse*, and a *universal*. Since, further, the disease may occur in combination with other diseases, or it may not, two kinds, a *complicated* and a *simple*, are recognised.

Description of Cases E and F.

I may here conveniently describe the case of sclerema neonatorum which I was able to investigate some years ago. I shall then record an instance of what I regard as oedema neonatorum which it was my good fortune to see at the same time. I place the two cases in juxtaposition, in order to bring out the resemblances and the differences which exist between

them, and also for the reason that many authors would doubtless regard them both as instances of sclerema, the former (Case E) being placed in the indurative variety, the latter (Case F) in the oedematous.

CASE E.

Clinical History.—On December 27th, 1885, an unmarried woman of 19 years of age was delivered, at the Edinburgh Maternity Hospital, of a premature male infant. The mother was a i.-para, and was in apparently perfect health. The labour was not a long one, the first stage lasting twelve hours, the second half an hour, and the third ten minutes. The vertex presented in the O.D.P. position. The child was small ; it weighed only 4 lbs. 12 oz., and had a length of 18 inches. The placenta, which appeared normal, weighed 1 lb. 3 oz., and the cord was 23 inches in length. The child, at the time of birth, had a premature appearance, and its breathing was with difficulty established, and, even when established, was shallow and weak. On the second day after birth it was noticed that the skin on the thighs, back, and buttocks was hard and tense, and had a dirty yellowish appearance. Pressure with the finger did not cause any pitting of the skin. The child took the breast very badly, and the cry was weak and whining. The indurated condition of the skin had, on the third day, spread to the neck, chest, and limbs, and was indeed almost universal. The body of the child felt cold to the touch, resembling the sensation given by contact with a half-frozen corpse. The child died on the evening of the third day of life, and through the kindness of Prof. A. R. Simpson I was enabled to make a post-mortem examination within twenty-four hours thereafter.

Morbid Anatomy: A. Macroscopic Appearances.—In the scleremic infant there were found, in addition to the conditions of the heart, vessels, and viscera peculiar to the new-born, the following pathological appearances :—The lungs were in a state of partial atelectasis, their posterior and lower portions being undistended with air ; the abdominal viscera, especially the spleen, liver, and kidneys, were markedly congested ; and the

brain and its membranes were also congested. The thymus gland appeared to be normal. It was in the skin and subcutaneous tissue that the most marked pathological changes were to be found. The skin over the back, shoulders, thighs, and to a less extent over nearly the whole of the body, was firm and tense, could not be pinched up between the fingers, and could not be made to pit on pressure. On making a section of the skin and subcutaneous tissue a sensation was conveyed to the hand like that got when bacon rind is cut; and, on looking at the cut surface, it was noted that the subcutaneous cellular tissue had a peculiar white glistening aspect, quite unlike the yellowish appearance of the subdermal adipose layer in the healthy infant. No serous fluid could be expressed from the cut surface, and to the naked eye the part did not appear to be in the least congested.

B. *Microscopic Appearances.*—In Plate I., Fig. 1, are represented the microscopic appearances of the skin and subcutaneous tissue from the back of the scleremic infant. The outstanding feature in this drawing is the presence of a large quantity of brightly-stained connective tissue, which forms a network, in the meshes of which lie the clumps of fat cells. This connective tissue is very abundant, and subdivides the subcutaneous adipose tissue into numerous patches of varying size. Not only are the bands of connective tissue increased in number, but they are also in many places much thicker than is normal. In the true skin are seen the roots of a few hairs, and one or two sweat glands with slightly convoluted ducts. Such were the appearances noted with a magnifying power of eighteen diameters. Under a higher power it was seen that the fat cells had in some cases lost all their fat, and that in no case was the normal amount of oil present. The nucleus in all was clearly visible, and there was often also a rim of protoplasm underlying the cell wall. The cells making up the bands of connective tissue could be clearly differentiated, and here and there were seen small vessels, surrounded by numbers of leucocytes, and pushing their way, as it were, into the clumps of fat cells. The papillæ were not well marked, and the outlines of the cells of the rete Malpighii were ill defined. The horny layer appeared to be

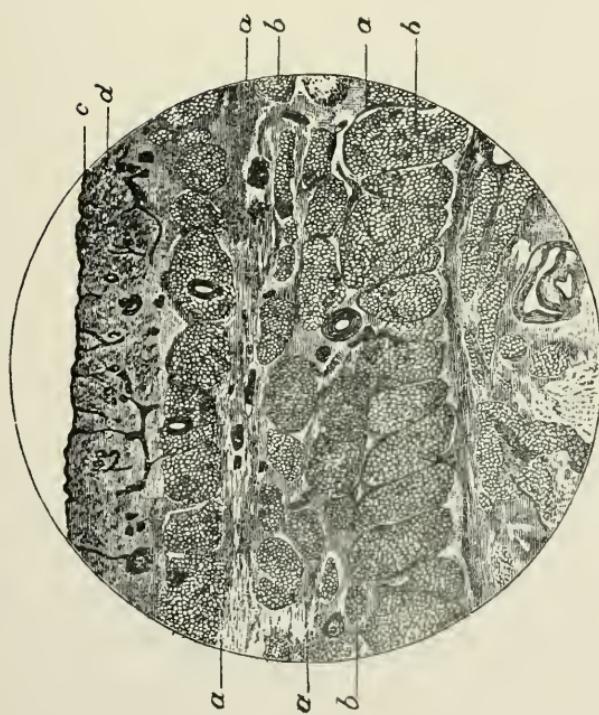


FIG. 1.—Solerema Neonatorum (Section of Skin of Back, $\times 18$). *a*, Bands of connective tissue; *b*, clumps of atrophied fat cells; *c*, epidermis; *d*, rete Malpighii.

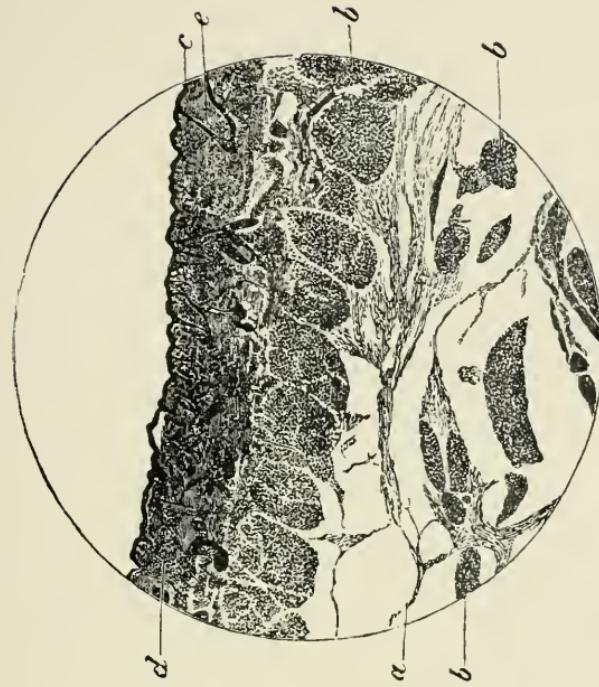


FIG. 2.—Edema Neonatorum (Section of Skin of Back, $\times 18$). *a*, Loose bands of connective tissue separated by serous fluid; *b*, groups of atrophied fat cells; *c*, cuticle; *d*, rete Malpighii; *e*, hair.

normal. The bloodvessels in the papillæ were very small, but those in the adipose tissue were relatively large, and had, as has been noted, a small cell infiltration surrounding them.

CASE F.

Clinical History.—On January 4th, 1886, a married iii.-para, of 31 years of age, was delivered, at the Edinburgh Maternity, of a premature male child weighing 6 lbs. 3 oz. The labour, which lasted for sixteen hours, was complex, for the mother was at the time suffering from acute bronchitis and had pneumonic patches in both lungs. The vertex presented in the O.L.A. position. The mother was removed to the Royal Infirmary fourteen days after labour, and there she made a tedious recovery. The child, which measured 18 inches in length and had a wasted appearance, developed in a few hours after birth a markedly œdematosus condition of the skin of the lower part of the trunk, of the external genital organs, and of the lower limbs, especially on their posterior aspect. The skin in these regions pitted readily on pressure, and had a bluish cyanotic appearance. The surface of the child's body felt moderately cold, but not nearly so cold as in Case E, and the limbs hung limp and inert. The child was put in the incubator, friction was applied, and stimulants were given; but notwithstanding these therapeutic measures, death ensued two days after birth. The child passed no urine. It may be stated that the placenta weighed 1 lb. 6 oz., and that the cord measured 19 inches in length. Neither placenta nor cord showed any abnormality.

Morbid Anatomy: A. Macroscopic Appearances.—The pathological appearances found in this case differed in some respects from those which have been described in the scleremic infant (Case E). Whilst the heart, liver, and spleen were normal, the kidneys showed on section an enormous degree of congestion, more especially of the cortex. The lungs, as in Case E, were in a state of partial atelectasis. The skin of the back, abdomen, external genitals, and legs was of a bluish colour, had a soft feeling, and pitted readily on pressure. When a section of the skin and subcutaneous tissue was made, a somewhat congested

surface was revealed, and a large quantity of watery serum drained away from the tissue.

B. *Microscopic Appearances.*—The microscopic appearance of a section of the skin of the back is represented in Plate I., Fig. 2. The masses of fat cells are seen to be held together in a loose manner by thin bands of connective tissue, and so loose is the connexion that in many places the fat cells have fallen out of the section. It was very difficult to obtain a satisfactory section of the subcutaneous tissue in this case, a circumstance which contrasted very forcibly with the ease with which perfect sections of the skin and underlying tissue were obtained in Case E. In the cedema section the tissue is opened out from the presence of the serous fluid, whilst in the sclerema case the tissue is indurated and the fat cells are held in position by the hypertrophied connective tissue framework. The fat cells in oedema, as in sclerema, are atrophied, and the appearances of the cuticle and rete Malpighii are similar in the two conditions. The difference appears to be in the subcutaneous adipose tissue layer.

The microscopic examination of the renal organs in Case F revealed the cause of the subcutaneous serous infiltration, for both kidneys were enormously engorged, the congestion being especially well marked in the cortex, and in both there was cloudy swelling of the cells of the tubules, and small cell infiltration of the Malpighian bodies. There was, therefore, evidence of the presence of tubular and glomerular nephritis, and this fact, taken in conjunction with the suppression of urine which was noted clinically, seems to show that in this case the disease and death were due to the kidney lesion. The infant died two days after birth, and as the renal changes found might quite well have arisen in that time, it was not necessary to believe that there had been foetal nephritis. The fact that the mother was suffering from bronchitis and pneumonia was, however, a noteworthy one.

CHAPTER II.

SCLEREMA NEONATORUM—Continued.

SYMPTOMATOLOGY; MORBID ANATOMY.

SYMPTOMATOLOGY.

IN considering the clinical aspects of sclerema neonatorum, a plan different from that employed in connexion with such a distinctly foetal disease as general dropsy must be adopted. In that malady the infant was either still-born, or lived only for such a short time that it had practically no clinical history apart from the mother ; its symptomatology was almost entirely intra-uterine, scarcely at all extra-uterine. It is quite otherwise with sclerema. This disease is rarely present at birth ; it develops, therefore, all its characteristic clinical features during the extra-uterine life of the infant affected by it, and has a symptomatology quite separate from that of the mother. Much greater space, therefore, is necessary for the consideration of its clinical characters.

Further, many of the contributions made to the study of sclerema contain not the records of individual cases, but the conclusions based upon the consideration of many observations not themselves set forth in detail. It is, therefore, impossible for me to treat this subject in the same manner as general dropsy and cystic elephantiasis were dealt with. With regard to the last-named diseases, it was possible to gather together the details of all the recorded cases, to compare these details, and so to form a conception of the characters of the maladies. One cannot consider sclerema neonatorum in the same exhaustive way for the reason stated above. Whilst I shall, therefore, treat the subject in a more general manner, I shall also occasionally mention certain outstanding peculiarities in individual cases.

It will be convenient in discussing the symptomatology of sclerema to consider, first, certain general and practically constant characters,—e.g., the fall in temperature and the diminution in body-weight; and, second, to take up in turn other clinical features which are more localised, less distinctive, and less commonly met with.

I. GENERAL AND PRACTICALLY CONSTANT SYMPTOMS.

1. *The Temperature.*

The lowering of the body-heat is the earliest, the most constant, and the most striking, as it is doubtless the most essential sign of sclerema neonatorum. The coldness of the surface of the skin was so evident in the first recorded specimen of the disease, that its observer (Usenbenzius) named it “fœtus . . . frigidus;” and almost every writer since has dwelt at some length upon this clinical feature. Soltmann (215) and G. Somma (150), who have given the most recent and the most comprehensive accounts of the malady, both regard the fall in the body-temperature as its most distinctive character.

The sensation given to the hand by the body of a scleremic infant has been described as one of marble-like or of icy coldness, and it has by some writers been aptly compared to that of a corpse in winter. I was much struck by the resemblance between the feeling given by the body of the specimen described in the preceding chapter and that of some infants that I had frozen for sectional purposes.

The information gained by the hand is fully confirmed and supplemented by that got from the use of the thermometer. In order to gauge the significance of the temperature readings in this disease, it is necessary to remember the variations in the body-heat which normally occur in the new-born infant.* At the moment of birth the temperature is about 37°25 C. (99° F. approx.). During the first half-hour of life it very distinctly

* For a detailed account of the temperature of the infant and its variations, the reader is referred to the Author's work, *An Introduction to the Diseases of Infancy*, p. 188. Edinburgh, 1891.

falls, reaching a lower level in feeble and premature infants than in healthy full-time children ; in the case of the former it may be as low as 33° C., whilst in that of the latter the lowest level reached has been placed at $36^{\circ}2$ C. (Andral), $36^{\circ}05$ (Roger), 36° (Baerensprung), or $35^{\circ}84$ (Eröss). This fall is a transient one, and in a few hours readings of 37° may be again registered. It is caused, in all probability, chiefly by the change in environment, and by the evaporation of the liquor amnii from the skin.

In scleremic patients the temperature falls far below that which is normal in even premature and weakly infants. Whilst such very low readings as $22^{\circ}06$ (Parrot and G. Somma), $22^{\circ}05$ and 22° (Roger), and $21^{\circ}08$ (Parrot), have been occasionally noted, the usual range in cases of sclerema is from 35° to 23° C., and most commonly the thermometer shows a body-heat of from 31° to 28° . The lowest reading noted by Soltmann (215) was $29^{\circ}8$ C. The temperature may be subnormal at the time of birth, as is seen in congenital cases ; and it would seem, from the observations of L. Somma (113) and Roger (93), that it begins to fall at a very early stage of the disease, and usually before the characteristic changes in the subcutaneous tissue have appeared.

With regard to the rate of body-cooling, G. Somma (150) states that two varieties of temperature-curve are met with. In one the fall is slowly progressive and comparatively slight, about 1° daily being lost, until a level of 28° or 27° is reached, morning and evening variations being absent ; in the other the cooling is rapidly progressive and greater, a fall of about 2° daily being registered, and a minimum of 27° , 25° , or even 24° attained in from three to four days. The first variety usually indicates partial sclerema of the oedematous type, whilst the second is associated with the diffuse and indurative form. Active heat-giving therapeutic measures may cause a slight and temporary elevation, and in the cases which recover a rise in the temperature occurs, as will be described later ; but, according to G. Somma, complications of an inflammatory nature are not able to raise the body-heat. All writers, however, do not agree with Somma on this last-named point,—thus Soltmann

(215) thinks that pneumonia may cause a rise in its early stage.

Parrot's observations (206) have shown that the skin temperature is the same as that in the body-cavities (mouth and anus); it may even be lower in the rectum than in the axilla, a circumstance which may be due to the application of hot bottles, etc., to the skin in the treatment of the disease.

2. *The State of Nutrition and Development.*

In many cases the subjects who afterwards develope sclerema appear at the time of birth poorly nourished; they have all the characters of prematurity, even although they may have been born at or quite near the full term. In other instances, as was pointed out by Underwood (219), Auvity (6), Soltmann (215), and others, they may come into the world fully developed and well-nourished, and the signs of denutrition appear later, either along with or immediately before the cutaneous manifestations of sclerema. The infant, if not weakly at birth, soon becomes so. The atrophic state shows itself in the small size, the shortness, the poor muscular development, but specially in the diminished weight of the infant. The weighing of scleremic infants comes then to be a diagnostic means of some importance, and the use of such an instrument as Sutils' portable infant-weigher makes it easy of application.* Normally a child at the moment of birth weighs about 3000 grms. (about 7½ lbs.); during the first two or three days of life it loses weight to the extent of 100 grms. (Bouchaud), 203 grms. (Gregory), or 222 grms. (Steiner), and thereafter it gains at the daily rate of about 25 grms. during the first and second months.† In the case of a scleremic infant the weight at birth is often less than 3000 grms. (it was only 4 lbs. 12 oz. in Case E); the initial loss is greater than usual; and the after-gain is either much less than normal or entirely absent, its place being taken by a slight loss.

It is a noteworthy fact, insisted upon by G. Somma, that both

* Ballantyne (J. W.), "Notes on a Portable Infant-Weigher," *Edin. Med. Journ.*, Oct. 1891.

† *Vide Author's Introduction to the Diseases of Infancy*, p. 194.

the loss in weight and the fall in temperature may be, and commonly are, present before the appearance of the subcutaneous tissue changes, so that they may in one sense be regarded as the prodromata of sclerema, and in another as its earliest manifestations.

3. *The State of the Integumentary System.*

The signs caused by changes in the skin and subcutaneous tissue are very evident and important, but they are not always uniform. The most constant of these signs, viz., the sensation of cold which the skin gives to the hand, has been already alluded to, and requires no further notice. A somewhat less constant symptom is rigidity of various parts of the body. This shows itself chiefly in the difficulty with which the limbs can be moved at the various joints. It has been stated (Dugès) that it may even be possible to seize the child by the head and hold it out like a piece of wood, but possibly this may have been due more to muscular contractions than to changes in the subcutaneous tissue. The skin cannot be pinched up in folds as it can in a healthy infant, neither can it and the subcutaneous tissue be made to glide over the underlying structures. The surface of the body becomes smooth, and the skin has a stretched appearance.

The consistence of the integumentary structures varies: in some cases there is a feeling of softness, the parts pitting easily on pressure; in others there is one of elastic hardness, the finger leaving no impression; and between these two extremes all possible grades exist. It was by many believed, and I myself shared in the belief, that it would be found that sclerema was always accompanied by induration of the subcutaneous tissue, and that it could thus be distinguished from what was called oedema neonatorum; but the observations of the Italian paediatricists and the occasional occurrence of the two conditions (indurative and oedematous) in the same patient—as pointed out by Denis (40), Valleix (221), Parrot (206), and G. Somma (150)—seem to show that a distinction founded upon this characteristic is not possible. G. Somma, who not infrequently found the combination of the two forms in the same subject, has used the

difference in the consistence of the parts as a means of dividing sclerema into three clinical varieties,—a truly sclerotic, an oedematous, and a mixed. In fact, he does not regard any one condition of the subcutaneous tissue as pathognomonic of sclerema. If his conclusions be correct, then the conflicting views of different observers with regard to pitting of the skin on pressure are easily explained. For whilst some authors (*e.g.*, Auvity) have denied the existence of this phenomenon, others have admitted it, and yet others (*e.g.*, Denis) have said that it occurs only when strong pressure is used. Some again, as, for example, Blanche, have held that the impression quickly disappears; whilst others (*e.g.*, Valleix) have stated that it is slowly effaced. It must now be admitted that digital pressure does not give a constant result, and cannot, therefore, be used for the diagnosis of sclerema, although it may be employed as a means of distinguishing its two chief varieties.

The parts of the body affected with sclerema are usually increased in size, and this change shows itself most markedly in the feet, thighs, legs, hands, and back, and to a less extent in the face and arms. It has sometimes been stated that so great is the increase that real deformity of the parts results, but possibly the cases in which this was noted were examples of true oedema due to cardiac or renal changes.

Authors have described in very different terms the colour of the skin in sclerema. The expressions yellowish-white, whitish-yellow, pallid, livid, leaden, dull-red, bluish-red, and brick-red, have all been in common use. G. Somma (150) thinks that these differences are due to variations in the seat of the disease, in its intensity, and in its extent, and to the presence or absence of complications; but whilst admitting all these factors, I believe that the colour of the skin depends also upon its tint at the time when the infant is attacked with sclerema, for it must be borne in mind that there are certain cutaneous colour changes which usually occur during the first few days of life. Physiological icterus neonatorum may be present, or pernicious icterus may be developed during the course of the disease, and in this way a yellow tint may be imparted to the skin. The drawing of the finger sharply along the skin causes a disappearance of the

reddish colour for a few seconds, its place being taken by a white or yellowish-white tint.

The skin is usually described as dry, perspiration being small in amount or absent, and it has been noted that if the normal neonatal process of desquamation has commenced, the supervention of sclerema arrests it. Further, it would seem that the cutaneous sensibility is either completely lost or greatly diminished in this disease, as is shown by the fact that pricking the skin with a pin or pinching it with the fingers usually produces no reflex movements. Either the sensory peripheral end-organs are insensitive, or from some fault in the nervous arc the sensation, although generated, is not transmitted. Delay in the separation of the umbilical cord is a symptom which has occasionally been noted, and may be mentioned here.

It is now necessary to consider the distribution of the above-mentioned cutaneous and subcutaneous manifestations of sclerema. They have sometimes been found to be nearly, if not quite, universal ; but usually these processes are at first limited to certain regions of the body. Commonly sclerema begins in the feet, in the cheeks, or in the hands, and spreads to other parts, viz., the legs, thighs, or arms. Sometimes the primary seat is the back, whence the disease extends to the pubic region, arms, and elsewhere. In the variety of the malady termed "sclerema diffuso" by L. Somma (113), it begins simultaneously in the face, feet, and hands. Verson (222) states that it always affects most markedly the regions in which adipose tissue abounds. Although a few exceptions have been noted, the skin of the front of the thorax usually enjoys an immunity from the scleremic process.

II. LOCALISED AND LESS CONSTANT SYMPTOMS.

The symptoms which have just been described may be regarded as the pathognomonic manifestations of sclerema ; but there are others, some of which are due to concurrent changes in certain of the internal organs, and some to morbid processes of the nature of complications. It is still uncertain to which of these two groups many symptoms ought to be referred ; they are, therefore, considered here together.

1. The Digestive System.

The signs of general weakness are found in the alimentary system, for at an early stage it is noticed that the infant sucks feebly or with difficulty ; but whilst this symptom is in most cases to be ascribed to muscular debility, in some it is due to muscular rigidity (pseudo-trismus). In addition to this there is usually difficulty in swallowing,—indeed, this act may be quite impossible.

The inspection of the mouth reveals the presence of parasitic stomatitis, at a comparatively early stage of the disease, according to Mildner (77) ; and it has been pointed out that this form of *muguet* is very intractable.

Constipation is met with at an early period in the development of the malady, and is caused, according to G. Somma (150), by the torpid state of the intestine,—for it, in common with the other organs, shows great functional depression. The same writer is of opinion that constipation, leading often to retention of the meconium, is rather more frequently met with in the oedematous and circumscribed variety of sclerema, whilst in the diffuse and indurative type a primary constipation is commonly followed by diarrhoea, which is often of a very grave kind, and is sometimes associated with vomiting. The diarrhoea is ascribed to gastro-enteritis, a very common complication. Various symptoms, e.g., jaundice, peritonitis, etc., due to changes in the liver, peritoneum, and mesenteric glands, may also be met with ; and pathological states of the umbilicus of an inflammatory nature (e.g., arteritis umbilicalis) have been described by Mildner (77), Soltmann (215), and others.

2. The Circulatory System.

One of the most evident symptoms of circulatory disturbance is found in the diminished rate of the pulse ; and this can be best ascertained by cardiac auscultation or by palpation over the anterior fontanelle, for the oedematous or indurative swelling of the subcutaneous tissue, the thinness of the vessel-walls, and the weakness of the heart's impulse, make the tactile examination of the radial or femoral pulse a matter of great difficulty.

Whilst the normal pulse-rate for the first days of life varies from 150 to 120 beats per minute,* in sclerema it falls to 90, 80, or even 70 per minute. Valleix (221) has placed the average between 60 and 78; but G. Somma (150) considers that at the beginning of the malady it is 85, and that it may rise to 90 or even 100 in the simple and partial form, whilst it may fall to 80 and even less in the diffuse and complicated variety of the disease.

Another character of the pulse in sclerema is its weakness. In the early stages it may be possible to feel the pulse at the wrist when it is small and sometimes irregular in strength and rate; but when the disease has reached its height it cannot be palpated there. In using the sphygmograph in cases of sclerema it would be necessary, in order to avoid error, to keep in mind the normal appearances of pulse-tracings in the new-born infant.

Auscultation reveals that the heart-sounds are scarcely audible, and it may also afford evidence of the presence of such complications as patency of the foramen ovale and ductus arteriosus, valvular lesions, and serous pericarditis. Auscultatory signs, as well as those gained from praecordial percussion and palpation, must, however, be always difficult of recognition in this disease.

3. *The Respiratory System.*

What would seem to be an almost constant symptom of sclerema is the slowing of the respiration. Whilst the normal neonatal rate is about 40 respirations per minute, in scleremic infants it soon falls to 35, 30, 25, 20, and even to 15 per minute. The higher rates indicate, according to G. Somma (150) the simple, partial, cedematous, and less severe forms of the disease; whilst the lower ones are associated with the complicated, diffuse, indurative, and grave varieties. The same writer has observed that the occurrence of pneumonia, a not infrequent complication, etards but slightly the diminution in the number of respiratory acts.

The respiration is irregular, sometimes painful, and is usually so superficial that no movement of the chest walls is visible. In some cases deep inspirations occur separated by intervals during which there is no sign of breathing.

* *Vide Author's Introduction to the Diseases of Infancy*, p. 162.

The cry in sclerema has been stated by Bierbaum (91) and others to be peculiar and even pathognomonic. It was compared by Dorfmüller (15) to the squeak of a young mouse. It has been described as distressful, veiled, smothered, whining, and squeaky (Soltmann). According to G. Somma (150) it may be hoarse and weak in the mild form, and sharp and hissing or whistling in the grave variety of the disease. The special timbre of the cry has been ascribed to oedema of the vocal cords and epiglottis.

Auscultation and percussion may reveal the presence of various pulmonary complications, such as pneumonia (lobar or lobular), atelectasis, congestion, and oedema, or of pleurisy with effusion. Cough, epistaxis, and the presence of sputum in the mouth have, therefore, been occasionally observed. Soltmann (215) states that pneumonia is the commonest complication of sclerema, and some of the early writers regarded it not as a complication but as an essential part of the disease.

4. The Urinary System.

Most authors go no further than to state that the urine is diminished in amount; but G. Somma (150) says that it is deficient in urates, uric acid, and, indeed, in all its principal physiological constituents. The secretion may disappear entirely in some of the more grave and complicated cases. It is almost colourless in many cases; but in those complicated by jaundice it is yellow, and leaves a distinct stain on the infant's linen. It is possible, as G. Somma suggests, that the retention in the system of certain excretory matters which ought to pass away in the urine may be the cause of some of the nervous phenomena of sclerema.

5. The Nervous System.

The nervous symptoms which have been described in cases of sclerema are not constant in character. They are more easily recognised in the indurative variety than in the oedematous, and are more marked in the widespread and complicated form than in the localised and simple. They usually take the form of convulsions, tonic and clonic, of the muscles of various parts of the

body, and near the close in fatal cases there may occur general eclamptic seizures. When the facial muscles are affected, all the appearances of trismus are present : the eyes are closed, chiefly by contraction of the orbicularis palpebrarum, and the jaws are clenched by the action of the masseters. When the muscles of the back are specially implicated, a condition not unlike opisthotonus is produced. As the muscular rigidity increases and becomes more widespread, the resemblance to tetanus is greatly increased.

With regard to the occurrence of paralytic phenomena, Angel Money (142) speaks very emphatically. He is of opinion that the want of movement in sclerema is due to paralysis, for in the three cases narrated by him faradisation failed to elicit muscular contractions.

In the grave cases, stupor, with torpor or coma, sometimes supervenes before the fatal issue, and it may then be difficult to decide whether the infant be living or dead. Coma, according to Valleix, is not frequently met with.

The symptoms of meningitis and other complications affecting the nervous system may also be present in some cases.

6. The Locomotory System.

The fact that the limbs of a scleremic infant are moved with difficulty or are quite immovable has already been mentioned ; but it may here be added that polyarthritis rheumatica is, according to Soltmann (215), an occasional complication.

7. The Integumentary System.

The constant symptoms met with in the integumentary system have been already referred to ; but it is necessary to state that certain complications may also occur. Cyanosis may be met with in the cases in which congenital cardiac anomalies exist ; jaundice is not uncommon ; erysipelas, pemphigus, and furunculosis sometimes occur ; and purpura and disseminated gangrene of the skin (Weickert, 109 ; Demme, 125) have also been observed. The cutaneous lesions of syphilis may complicate sclerema.

MORBID ANATOMY.

In the description of the pathological changes met with in sclerema, several methods have been employed. G. Somma (150), for instance, has divided them into two groups,—the first containing the cutaneous lesions, which are constant, essential, and permanent; whilst the second includes the visceral changes, which are occasional, accidental, and transitory. There are, however, certain morbid alterations in the skin which are neither constant nor essential, and it may be that some of the visceral lesions are of more importance than has been thought. Other writers—*e.g.*, Depaul (122)—have described the morbid anatomy according to the system of organs affected; and this plan I intend to adopt, considering first the most important alterations, viz., those met with in the integumentary system.

I. *The Integumentary System.*

A. *Macroscopic Appearances.*—Certain of the changes in the skin, discovered by palpation and inspection, have been considered along with the clinical phenomena, so that it is now necessary to discuss only those met with on dissection.

The *epidermis* has usually a normal appearance, but the *dermis* shows alterations which may be of two kinds. Sometimes there are the manifest signs of congestion: dark-coloured blood flows from an incision or prick, and the tissue is soft and loose. In other instances the dermis has an anaemic, not a congested appearance; it is firm to the touch, whitish-yellow in colour, and no blood flows from it when cut into. In the *subcutaneous tissue* similar changes are met with. In one group of cases the subdermal layer is congested and infiltrated with serum, which is transparent, of a yellow colour, coagulable by heat, for it is rich in albumen, and does not give the reactions of bile-pigment. There is no pus found, save in the cases in which erysipelas or furunculosis is present as a complication. In another set of cases—the indurative—the subcutaneous tissue is free from oedema, is yellowish-white in colour, of a firm or hard consistence, and contains, closely packed together, clumps

of fat. In nearly all the cases a transparent gelatinous mass, from which pressure expels some serum, is found, lying closely upon the subjacent aponeurosis (G. Somma). In the *intermuscular connective tissue* there is probably always some signs of oedema, more marked in the oedematous variety, less so in the indurative. With regard to the *capillaries* of the skin, sometimes they are greatly distended with blood, at other times they are obliterated at places as well (Bouchut). Little or nothing is known concerning the *lymphatics*, although Virchow regards the disease as essentially a lymphatic oedema.

B. Microscopic Appearances.—In the indurative variety of sclerema the most important change is found in a great increase in the connective tissue of the subcutaneous layer. It forms a close network, in the meshes of which lie the fat corpuscles in small clumps. With a higher power a small cell infiltration of the dermis and subdermal layer can be seen, and this probably constitutes the primary change in the scleremic process. According to Pavone (152), this infiltration affects also the rete Malpighii of the epidermis as well as the dermis. The formation of the connective-tissue network above described is no doubt a secondary change. The fat cells show a very distinct nucleus, and this is due to the almost complete disappearance of the oil from their interior.

In the oedematous form the connective-tissue network is much looser, and in mounting microscopic sections the clumps of adipose tissue often fall out. There is probably the same parvicellular infiltration as in the indurative variety of sclerema. The fat cells have also lost most of their contained oil.

In both forms the stratum corneum of the epidermis appears normal, whilst the cells of the rete Malpighii have blurred outlines. The bloodvessels supplying the dermal papillæ have been described as narrow and slender, whilst those in the adipose tissue layer are larger. Obliteration of the sudoriparous glands has also been noted.

Besides the lesions above noted, which may be regarded as the characteristic ones in sclerema, there may be met with certain changes which are accidental, and are caused by erysipelas, pemphigus, and syphilis. Hæmorrhage into the skin,

constituting purpura, may also be found, as well as the signs of cutaneous gangrene.

2. The Digestive System.

Various morbid appearances have been described in connexion with the viscera of the alimentary system, but most of these are doubtless accidental phenomena. For instance, Léger (46) was of opinion that in sclerema the intestinal canal was always shorter than normal. He stated that in a healthy infant the intestines should measure 10 feet, and in one that has died from enteritis, 14 or 15 feet; but that in a scleremic infant it would be found to vary in length from $4\frac{1}{2}$ to 10 feet. In 55 per cent. of the scleremic cases he found a length varying from $6\frac{1}{2}$ to $8\frac{1}{2}$ feet, and he considered that this shortness of the intestine was of great etiological importance. Most writers, from Billard to G. Somma, have not, however, been able to confirm Léger's observations.

Most of the digestive organs have been found to be markedly congested,—thus red patches have been described on the gastric and intestinal mucous membrane; and it has been noted that the liver and spleen are commonly enlarged, softer in consistency, and darker in colour than is normal. Little food, but much gas, has been found in the intestine, although sometimes the colon has contained a considerable quantity of meconium. In the stomach and intestine, ulcers and gangrenous patches have been described; and enlargement and caseous degeneration of the mesenteric glands have been seen in some instances. Amongst other morbid changes, the presence of serous fluid in the peritoneal cavity, and the signs of arteritis and phlebitis umbilicalis, have been observed.

3. The Circulatory System.

In the cavities of the heart and in the aorta and large vessels there is usually much dark-coloured blood; and Valleix (65) has described polypus-like masses in the right auricle, which are probably fibrinous deposits. The walls of the heart are soft, and there may be fatty degeneration of the myocardium. Sometimes the heart is hypertrophied, and it is not uncommon

to find delayed closure of the foramen ovale, ductus arteriosus, and ductus venosus Arantii. Fluid is often found in the pericardial sac ; it is sometimes serous and sometimes sanguinolent. The pericardium itself may show ecchymoses on its walls. Closure of the capillaries (Clementovsky) and obliteration of the thoracic duct and its branches have occasionally been noted. G. Somma (150), however, states that the lymphatics are usually normal.

4. The Respiratory System.

There is general congestion of the respiratory organs. This shows itself in the mucous membrane of the larynx and trachea, and there may also be an œdematous swelling of the epiglottis and vocal cords. There is usually pulmonary congestion, along with more or less widespread atelectasis. Pneumonia, either croupous or catarrhal, is frequently met with ; some of the older authors (*e.g.*, Hulme) considered it to be constant, and attached to it great etiological importance. Hæmorrhages into the lungs and pulmonary gangrene have also been seen. Ecchymoses occur on the walls of the pleural sacs, and clear yellow fluid in their interior. Pleurisy is a rare complication.

5. The Urinary System.

Pathological changes in the kidneys are not commonly met with, but there is often considerable renal congestion. Sometimes the mucous membrane of the bladder shows ecchymotic spots.

6. The Nervous System.

The same congestion which is so evident in the digestive, respiratory, and urinary organs is met with also in the brain and its membranes. There is nearly constantly meningeal congestion, and this affects chiefly the pia mater. Small hæmorrhagic foci are sometimes seen in the brain, which may also be anaemic or œdematous. The cranial sinuses contain much blood, and intra-cranial cephalhæmatomata are sometimes met with. The cerebral ventricles are often distended with serum, and an increase in the amount of fluid in the spinal

canal has also been noted. The changes due to meningitis, a not uncommon complication, have sometimes been observed.

It will have been gathered from this review of the morbid anatomy in sclerema that the most constant changes are those due to a disturbance of the circulation, and that in the case of the internal organs this disturbance is of a congestive kind ; whilst as regards the skin and subcutaneous tissue, it may be either anaemic or congestive. It is, further, interesting to find that Pavone (152) is of the belief, from his microscopic investigations, that the same primary change, viz., a small cell infiltration, underlies both the varieties of pathological alteration in the integumentary system. The morbid appearances which are not constant are chiefly of an inflammatory nature. The bearing of these facts on the pathogenesis will be discussed later.

CHAPTER III.

SCLEREMA NEONATORUM—Continued.

ETIOLOGY; PATHOGENESIS.

IT is now necessary to consider what conditions have been regarded as etiological factors in the production of sclerema, and what theories have been advanced to explain the nature of the malady. With regard to its etiology there is a considerable degree of unanimity amongst the various authorities ; but in all matters relating to its pathogenesis there has been an astounding diversity of opinion. The etiological factors that are of prime importance have been almost decisively agreed upon ; but as to the manner in which they act on the infantile organism much confusion exists.

ETIOLOGY.

In discussing the etiology of sclerema authors have differed greatly in their views concerning the predisposing and the exciting causes of the disease. Some have considered bad hygienic surroundings as predisposing factors, others have regarded them as determining causes ; and many similar examples might be given. I shall, in this place, simply consider the various conditions which have been looked upon as etiological, and shall thereafter attempt to classify them according to their importance and mode of action.

I. *Age.*

One of the conditions which all authors have regarded as of prime importance in the causation of sclerema is the age of the patient. The disease is emphatically one which occurs during the earliest days of life : it is a neonatal malady. It is not, however, limited absolutely to this period of life, for exceptions are met with in two directions : it may occur after the first

month, and it may also be developed before birth. Usually sclerema makes its appearance at some time in the first three weeks of life, and most commonly during the first five days. From the second to the fourth day is the age of predilection. In more than two hundred cases observed by L. Somma (113) the disease was developed in practically every instance between the third and the sixth day of life. Its early signs have, however, been noticed within a few hours of birth in certain cases—Heyfelder (192) and others,—and in other instances the disease has first appeared in the second, third (119), and even fourth week of extra-uterine life. Its appearance at a later date must be regarded as quite exceptional, and it may be doubted whether some of the examples reported at one, two, or three years of age have been sclerema at all. Crocker (171), at any rate, is of opinion than Isambert's case (100) at thirteen months was really scleroderma and not sclerema. MacAndrew's patient (54), who first showed signs of the disease at sixteen months, Barlow's (115), who developed it at three and a half years, and Angel Money's three cases (142) at four and a half, eight, and nine months respectively, seem, however, to have been genuine examples of sclerema.

With regard to the foetal cases, whilst most authors say that the disease is sometimes present at birth, and while some go as far as to state that it is generally congenital—"meistentheils angeboren," Feiler (182),—very few record individual cases illustrating this. Still a sufficient number of observations has been made to justify the statement that sclerema is occasionally truly congenital. The case reported by Usenbenzius (1) was one of this kind, and Graetzer (187), writing in 1837, was able to collect together some ten or twelve cases. According to this writer, the six months' fœtus described by Deutschberg (33) suffered from congenital sclerema, and the specimens of false ascites (*la fausse ascite*) noted by Dugès and others, were also examples of that disease. Two of the cases of sclerema reported by Denis (172) exhibited the morbid process at the time of birth, and Alibert (153) and Orfila (205) reported other illustrative examples. Congenital cases have also been described by Horst (67), Hervieux (87), Demme (125), and Soltmann

(215); and it would seem from the statements made by Underwood (219), Gardien (185), and Billard (163), that they also had noticed the occurrence of the malady in the infant at birth. Soltmann (215) mentions a congenital case reported by Suckling, but does not give the reference.

From what has been said it may be concluded that whilst the cutaneous manifestations of sclerema are usually not evident till some days or hours after birth, there are, nevertheless, exceptions to this general rule. Further, when it is remembered that a sub-normal temperature is an earlier symptom than either œdema or induration of the subcutaneous tissue, it may reasonably be believed that many other instances are truly congenital as regards the occurrence of that symptom. Again, it must not be forgotten that many scleremic infants have been born prematurely, and that, therefore, although the disease truly attacked them in extra-uterine life, they were yet only seven, eight, or nine months' foetuses in all respects save environment. It is possible, then, that sclerema may be as much a foetal as a neonatal disease.

2. *Sex.*

It would seem that the sex of the infant has little or no effect upon the development of sclerema. Heyfelder (192) stated that upon the whole as many boys as girls suffer from it, and noted some epochs in which the male sex was more liable, and others in which the disease more commonly attacked females. L. Somma (113), in a series of two hundred and twelve cases found one hundred and thirteen males and ninety-nine females; but in another series he noted that there was a preponderance of the latter sex. According to G. Somma (150), Blasi observed the disease fifty-six times in male and thirty-seven times in female infants.

3. *Constitution.*

It must, I think, be conceded that most of the infants that are attacked by sclerema show at the time of birth a certain amount of congenital debility, are poorly nourished, are below the normal as regards both size and weight, and are particularly susceptible

to all morbid influences. At the same time it must be remembered that all authors are not agreed upon this point ; for, whilst some, such as Billard (163), Valleix (221), Vogel (223), Marcacci (107), Roger (74), West (226), Schwimmer (213), and G. Somma (150) are of the opinion stated above, others, such as Underwood (219), Auvity (6), Hulme (8), Soltmann (215), Clementovsky (111), and Blasi (150), believe that infants with the best constitutions may equally be affected with sclerema. When it is borne in mind that the weight of such infants is nearly always much below the normal, it is difficult to imagine that their organic constitution can be anything but below par. Congenital debility is probably one of the most important etiological factors.

4. *Congenital Morbid States.*

Whilst some writers have dwelt upon the importance of congenital debility, others have insisted that definite antenatal morbid states are present, and that they markedly predispose such infants to the development of sclerema. Demme (125), for instance, insists upon the etiological importance of the presence of foetal myocarditis ; Letourneau (198) and West (226) look upon pulmonary conditions, especially of the nature of atelectasis, as causes ; and Clementovsky (111), Soltmann (215), Kutsch (26), and others, have ascribed sclerema to congenital anomalies of the blood and lymph vessels. Congenital anaemia has been mentioned by Löschner (90), and antenatal syphilis by Doublet (4), Feiler (182), and Gölis (20). Whilst it may be admitted that such morbid states may act as causes predisposing to sclerema, it is, I think, wrong to regard this disease only as one of their symptoms.

The four groups which have just been considered have been regarded by G. Somma (150) as containing the intrinsic predisposing causes of sclerema, and of them all he has given the greatest etiological importance to congenital debility. Too little attention has, however, been paid to the possible causal factors which may be found in the state of health and surroundings of the parents of scleremic infants. The existence of any paternal influence can neither be affirmed nor denied, for practically nothing is known about it, although Demme (125) states that in

the case observed by him both the parents were in poor health from the use of bad food and from over-indulgence in alcohol. I shall, therefore, deal only with possible maternal causes.

5. *State of the Mother's Health, etc.*

Since most of the recorded cases of sclerema have occurred in foundling asylums, little information concerning the maternal history has been elicited; but the very fact that the infants affected have been found in such institutions proves that the mothers must have been in very poor or in distressing circumstances, and it may also be surmised that in many cases they may have been the victims of syphilitic infection. The same remarks apply to the cases which are met with in maternity hospitals.

When definite information is forthcoming with regard to the maternal history, it is usually found that scleremic infants are the offspring of young, unmarried, primiparous women, who have been living in such circumstances of poverty and wretchedness as are unknown to their happier married sisters. It must be admitted that from the time of conception the foetus is being nourished within an organism depressed by all the conditions associated with seduction, or with a life of shame and debauchery. Northrup (148), a recent writer, insists upon the etiological importance of poverty and squalor in the mother; and Reydellet (208), an older authority, recognises the same causes when he says: "La misère, les souffrances de la mère, et mille autres causes de cette nature, en altérant le fruit de la conception, ne sont-elles pas le principe du mal?" Graetzer (187) is of opinion that struma or syphilis in the mother during her pregnancy may, by depreciating the nourishment supplied to the foetus, lead to the development of sclerema.

It has often been noted that the pregnancy that results in the birth of a scleremic infant ends prematurely at the seventh or eighth month; and according to Namias (133), Chiara is of opinion that prematurity is the primary and necessary factor in the development of the disease. It has also occasionally been found that the gestation is a plural one. Northrup's case (148) was one of twins, as was also one of the examples reported

by Namias (133); whilst Horst (67) put on record an instance of sclerema in triplets. Carminati (35) states that the disease is specially common in twins. These peculiarities in the gestations that result in the birth of scleremic infants, whilst they may have no direct effect in the production of the disease, must certainly induce congenital debility in the offspring, and so act as indirect causes. Some of the older authors were of opinion that certain circumstances occurring during the pregnancy were the immediate factors in the development of sclerema. Usenbenzius (1), for instance, ascribed the malady in the case observed by him to the fact that the mother when pregnant had spent much of her time in the contemplation of the statues in the churches. Such theories may be neglected; but it is doubtful whether there may not be some truth in the statement made by Villebrun (quoted by Meissner in his "Forschungen," 202) to the effect that the liquor amnii is so modified as to exercise a tanning influence upon the body of the fœtus in utero. At any rate, it would be well in such cases to examine the foetal annexa.

The confinement does not seem to have been often abnormal in the reported instances of sclerema, and the puerperium was usually perfectly favourable in its course. One fact which may be of some importance has been occasionally noted; and that is, that sometimes the same mother will give birth to several scleremic infants in succession; thus, in Angel Money's paper (142) it was noted that three sisters were affected, whilst the only other infant, a male, escaped.

We may now pass on to consider some of those causes which have been termed by G. Somma (150) extrinsic and predisposing, and which consist in peculiarities in the environment of the infant after birth.

6. Climate and Geographical Position.

It has usually been stated that sclerema is more often met with in countries with a temperate climate than in the tropics or in northern regions; and it has been added that the climate that most markedly predisposes is a variable one, subject to frequent and sudden vicissitudes of temperature. It may at

any rate be stated definitely that whilst the disease has been met with and described in Great Britain, Germany, Austria, Russia, Spain, Switzerland, Denmark, and the United States, it is infinitely more common in France and Italy. In fact, it is an extremely rare malady in the first-named countries. In Paris, towards the beginning of the present century, the disease occurred with alarming frequency. Tenon (*Mémoires sur les hôpitaux de Paris*, 1816) states that of the 6000 infants cared for annually in the Parisian foundling hospitals, 600 died of sclerema ; and Baron and Breschet report the occurrence of 350 cases of the disease in 1822, and 400 in 1823.

With regard to Italy, Loder (21) affirms that Bruni saw about one hundred cases yearly in the Foundling Hospital at Florence; Liberali (29) made seventy autopsies on scleremic infants in one year ; and L. Somma (113) gave statistics of two hundred and twelve cases. G. Somma (150) has also pointed out that the disease is most common in the southern provinces, and especially in Rome and Naples. The frequency with which sclerema is met with in France and Italy may, of course, be due to other causes than climate ; but the fact is nevertheless a most striking one.

7. *Race.*

The rarity of the disease in Germany, Austria, and Great Britain, and its common occurrence in France and Italy, seem to point rather to a *racial* than to a climatic causal influence. It may be that the Latin race is more liable to sclerema than are Teutonic and Sclavonian peoples. The fact that little has been written on the subject by Spanish authors does not necessarily prove that it is rare in Spain.

8. *Season of the Year.*

It has been admitted by all observers (with one exception) that sclerema is more common in the winter than in the summer months. Liberali (29) never saw it in June, July, August, September, or October ; Elsässer (81) noted it most frequently in November and December ; L. Somma (113) found that it occurred commonly in winter and very rarely in summer ; and G.

Somma (150) observed it most frequently in December, January, and February, and saw only a few cases in the summer months, and then only when the temperature happened to be more variable than usual.

The following statistics, taken from the works of Valleix (79 and 221) and Billard (163), are very suggestive :—

Month.	<i>No. of Cases.</i>	
	Valleix.	Billard.
January,	50	15
February,	37	15
March,	56	16
April,	49	18
May,	22	22
June,	19	3
July,	12	4
August,	0	14
September,	3	10
October,	24	16
November,	23	29
December,	15	15
	—	—
	310	177

Only one writer of note, Palletta (38), observed that sclerema occurred with greater relative frequency in the warm months of summer and autumn than in winter. It is possible that some other causal influence was at work in Palletta's cases, and that this counteracted the effect produced by the season of the year. It may, however, be stated that, as a general rule, sclerema occurs most frequently in the cold and damp months.

9. *Hygiene.*

As has been already pointed out, it is the infants of unmarried women of the poorer classes who suffer most from sclerema. It is rare to meet with a case among the well-to-do and in private practice. It follows that the hygienic surroundings

of such infants will usually be found to be of a most unsatisfactory nature. They are born in cold, damp, imperfectly-lighted and badly-ventilated houses; are insufficiently clothed; and are, in all probability, seldom or never bathed. All these circumstances must tend to increase their liability to be attacked by sclerema; but I do not think the facts warrant us in concluding that they are anything more than predisposing causes. An author—quoted by Henke (189)—states that the disease is immediately due to the putrefaction of the vernix caseosa left upon the infant's skin from imperfect cleansing; but there is not sufficient evidence to make this hypothesis tenable.

10. *Diet.*

Since most scleremic infants are met with in foundling hospitals, it follows that their feeding must, in the majority of cases, be artificial. They run the extra risk involved in rearing on the bottle. In the few cases in which the mother's milk is available for their nutrition, it is probable that its quantity has been diminished and its quality altered by the conditions under which the woman has lived. The feeding of the infant with insufficient or improper food must be regarded as yet another predisposing factor in the etiology of sclerema.

11. *Cold.*

Nearly every author has regarded the action of cold upon the body of the infant as an important cause of sclerema, and G. Somma (150) believes that it is the only *determining* factor in its etiology, giving as his reason for this belief the frequent occurrence of the disease in the cold months of the year, in countries in which the temperature shows numerous and sudden changes, and in foundling hospitals and among the poor. In order to explain away the facts that the disease is rarely met with in such a cold country as Russia and in such a variable climate as that of Great Britain, and that it is sometimes met with in summer, this writer goes on to state that the atmospheric cold is not the only chilling influence that may act upon the body of the infant. He points out that cutaneous refrigeration

may also be produced by the use of cold baths and by insufficient clothing of the child. Further, whilst he gives great etiological importance to chilling of the external surface of the body, he is forced to admit the existence of a predisposing cause (congenital debility) in the case of the infants who develop sclerema. The greatest difficulty that I find in accepting chilling of the body as the only determining cause of sclerema consists in the occasional occurrence of the disease before birth. From what is known of intra-uterine physiology it seems almost impossible to conceive that the temperature of the surrounding parts can be so much diminished as to cause a chilling of the foetal organism so great as to produce the disease. At the same time there seems to be no doubt that cutaneous refrigeration is in most cases a very important factor in the causation of sclerema.

12. *Contagion, Dentition, etc.*

Only a passing notice may be given to one or two other conditions which have been supposed to excite the disease. For instance, Bard (23) suggested that dentition might be a causal factor ; but then it is well known that in the vast majority of cases sclerema develops long before teething commences. When it is borne in mind that history records the occurrence of large numbers of cases of this malady at the same time and in the same hospital, it seems surprising that contagion has not been frequently advanced as a possible cause. Mignot (204), however, seems to have held some such theory, and Baginsky (156) suspects the existence of an infectious influence leading, in the case of delicate infants, to an anomaly of the walls of the smallest bloodvessels. There is as yet no sufficient evidence of this, but Semet (152A) seems to regard the theory with favour.

It has also been stated that social position is an etiological factor in the production of sclerema : for whilst the infants of the poorer classes are liable to it, those of the well-to-do escape ; but it is evidently the environment and not the mere social status that is the determining agent in these cases. Finally, prolonged decubitus in the horizontal posture has been emphasized by Hervieux (83A).

Such are the alleged causal factors that have been noted by

various authors, and it will be seen that they can be arranged in two groups: in the first are those external to the infant or foetus, viz., the climatic, hygienic, and dietetic causes, and the state of health of the mother during pregnancy; and in the second group are the intrinsic conditions, viz., the age, constitution, and state of health of the infant itself. They may also be classified as predisposing and determining, and whilst G. Somma regards cutaneous refrigeration as the only cause in the second class, and all the others as belonging to the first, I feel inclined to believe that there may be more than one determining factor, and that the most important cause is the existence of a congenital morbid tendency which, when acted upon by various external agents, leads to the development of sclerema.

PATHOGENESIS.

The etiology of sclerema has been discussed, and we are now in a position to consider the various theories which have been advanced to explain the ultimate nature of the morbid process and the mode of action of the causal factors. The extraordinary diversity of these theories is in part to be accounted for by the fact that the writers were sometimes treating of examples of diseases other than sclerema; their opinions may have been quite correct with regard to the cases they were describing, but then these cases were not instances of sclerema. The theories may be arranged in three groups: those in the first may be called the *diathetic or dyscrasic*, and according to them the disease is simply one of the manifestations of syphilis or rheumatism; those in the second may be termed the *inflammatory*, the malady being regarded as due to an inflammatory state of the skin and subcutaneous tissue; and those in the third are the *œdematosus*. With regard to the last group there are several sub-groups of theories, according as the cause is supposed to lie in the circulatory, hæmopoietic, respiratory, alimentary, or nervous systems.

1. *The Syphilitic Theory.*

Doublet (4) was the first to suggest that sclerema neonatorum was a manifestation of syphilis; Feiler (182) seems also to have

been of this opinion ; and Gölis (20), whilst he thought that the sclerema of the French writers was an aborted erysipelas, held that the skin-bound disease of the English physicians was a chronic syphilitic process. This theory has now been generally abandoned ; for whilst it is true that sclerema is sometimes complicated by the presence of congenital syphilis, and whilst it is also certain that syphilitic infants occasionally show some degree of subcutaneous oedema, sclerema occurs most frequently in babes that are quite free from the venereal taint.

2. The Rheumatic Theory.

A passing reference may here be made to another diathetic theory, that advanced by Barthez and Rilliet (158). These writers considered that sclerema was rheumatismal in nature. It is true that in one of Demme's cases (125) acute polyarthritis rheumatica occurred ; but this was no doubt simply a complication.

3. The Inflammatory Theory.

The theory that sclerema was inflammatory in nature was held by many authors, especially amongst the Germans. Some were of opinion that it was a form of erysipelas, and others believed that it was simple inflammation.

(a) The Erysipelatous Theory.

Underwood (219), in the first edition of his work, stated that sclerema was an anomalous form of erysipelas ; but he departed from this view in later issues. Lodemann (17), Gölis (20), and Zimmermann (39) thought that the sclerema of the French writers was erysipelas neonatorum, possibly an aborted form. Fischer (18) held the opinion that it was the same kind of disease as erysipelas ; and Henke (189), although he recognised a difference between sclerema and erysipelas, thought that both these diseases, and also icterus neonatorum, were closely allied, and that possibly the development of one or other might be due to some difference in the external conditions. Amongst recent writers Clementovsky (111) seems to have recognised some bond of union between this disease and erysipelas. Sodaffsky (43),

however, and most of the other observers, rejected the theory of an identity between sclerema and erysipelas. There seems to be no doubt that some of the cases described as sclerema were examples of erysipelas, and the latter disease may possibly occur sometimes as a complication of the former; but there is no evidence to show that sclerema is erysipelatous in its nature.

(b) *The Simple Inflammatory Theory.*

Carminati (35) regarded sclerema as an inflammatory process which might manifest itself clinically either in a sthenic or in an asthenic form; and Denis (40) thought that it was true inflammation of the subcutaneous cellular tissue associated with various internal maladies, and more especially with gastro-enteritis,—that it was, to put it shortly, an entero-cellular phlogosis. Mildner (77) was of the opinion that underlying the ultimate manifestations of sclerema was a primary inflammatory process affecting the subcutaneous cellular tissue and derma, and sometimes also the adipose layer, and he founded his belief upon the results of autopsies in which inflammatory conditions of various organs and of the umbilical vessels were discovered. Possibly he may have been describing cases of puerperal fever in the new-born. Hennig (190), who considered chiefly the œdematos variety of sclerema, thought that it was at first due to passive congestion of the general integument, and that this became later a true inflammatory process as a consequence of the weakness of the organism and of the delicate state of its cutaneous tissues. According to G. Somma (150), Pastorella was of opinion that the malady was an inflammatory affection of the peripheral lymphatic vessels.

All these theories have now, however, been generally abandoned, and most writers have come to the conclusion that sclerema is an œdematos condition; but the cause of the œdema has been very variously stated, some having looked for it in morbid states of the heart, blood, and lungs, and others in lesions of the digestive organs or nervous system.

4. *The Cardio-Vascular Theory.*

Both Andry (5) and Auvity (6) held that the action of cold

interrupted the insensible transpiration from the skin and slowed the circulation, and so led to oedema. Jahn (194) thought that sclerema neonatorum was the same as *morbus coeruleus*, and since his time several writers have looked to congenital anomalies of the heart for an explanation of the disease. Breschet, for example, cited patency of the foramen ovale, and others advanced as causes an open state of the ductus arteriosus or cardiac valvular anomalies. Demme (125) considers that the starting-point of the disease is a fatty degeneration of the myocardium (especially of the right ventricle) which by weakening the muscular energy of the heart, leads to slowing and obstruction of the circulation.

It would seem that Billard (163) considered the process as an oedema due chiefly to circulatory disturbances. He gave as predisposing causes (1) natural feebleness in the infant, (2) a state of general and congenital plethora, (3) excess of venous blood in the tissues, and (4) the state of dryness of the skin preceding the neonatal exfoliation of the cuticle; while he looked for direct causes in (1) an obstacle to the circulation of the blood from excess of the same in the vascular system, (2) accumulation of the blood in the cellular tissue with consequent serous infiltration, and in (3) the action on the skin of external agents. Bouchut (164), also, thought the disease was an oedema due to a disturbance in the circulation in the skin; but as G. Somma has pointed out, he did not attempt to explain the cause of this hydraulic perturbation. Carus (25) thought that sclerema was due to a lowering of vital activity in general, affecting specially the cutaneous system, and leading to a condition comparable to marasmus senilis and to the so-called "dead fingers." Legroux (87A) considered that it was a syncopal asphyxia from muscular inertia.

Soltmann (215), a recent writer on the subject, has regarded sclerema (*sklerödem*) as a disease due to a morbid condition of the vitality of the blood (*eine krankhafte Disposition des Blutlebens*) and of the vessel walls, leading to oedematous infiltration of the subcutaneous tissue from porosity of the capillary vessels (*Lockerheit der Capillargefässer*).

Dumas (141), in a long paper, sought to prove that oedema of

the new-born was only a symptom of phlegmatia alba dolens developed in the early days of life, the thrombus forming most frequently in the vena cava inferior, and being specially predisposed to by an imperfect establishment of respiration. The difference between the symptoms of phlegmatia alba dolens in the infant and in adult life he regards as due to the special physiological conditions existing in the early days of life. Dumas' theory is not new. Lodeman (17) as long ago as 1810 pointed out the resemblance between sclerema and phlegmatia alba dolens.

It may here be stated that some writers have looked to the lymphatic system for the causal lesion of sclerema. Virchow regards it as a lymphatic oedema, and believes that it is closely allied to elephantiasis. Kutsch (26), also, writing in the early part of this century, regarded it as due to congenital anomalies of the lymph-vascular system. Heller's case, however, in which there was obliteration of the thoracic duct and its branches, seems to have been an instance of scleroderma and not of sclerema. Rigal (104A) advanced a theory in which he stated that the disease was due to insufficient action both of the heart and of the lymphatic vessels. The great objection to all these theories lies in the fact that many observers have demonstrated an intact state of the lymph vessels in sclerema.

5. *The Pulmonary Theory.*

From the frequent association of sclerema and pneumonia Hulme (8) was led to believe that the former was a symptom of the latter, and it would seem that Dugès (176A) and Troccon (22) at first were of the same opinion, but afterwards finding that pneumonic changes were far from constant, abandoned the theory.

Ritter (209), recognising that pneumonia was not always present to account for the imperviousness of the pulmonary tissue, looked for other causes in atelectasis and pleurisy, and regarded the disease as the expression of both a local and a general disorder due to partial condensation of the structure of the lungs. Palletta (38) had in 1823 advanced the theory that a flaccid state of the lungs (especially of the right) arrested the

circulation and so produced sclerema ; but in a second communication (49) he held that cold hindered the expansion of the thorax and dilatation of the lungs, contracted the pulmonary vessels and alveoli, and arrested the circulation (already wanting in force) in the vascular system and in the viscera. Frank (11), also, was of the opinion that the regular accomplishment of the process of respiration was hindered in various ways (by the action of cold upon a weakly infant, insufficiently fed, and living in an impure atmosphere), and so the oxygenation of the blood and the development of animal heat were interrupted, and the fluid tissues passed into a solid state.

6. The Cardio-Pulmonary Theory.

Some authors, recognising that neither the cardiac nor the pulmonary theory was sufficient in itself, looked for the pathogenesis of the malady in a combination of the two. Sclerema, according to this combination theory, was a symptomatic œdema due to an impeded circulation in the heart, lungs, and vessels. Heyfelder (192), for instance, pointed out that the process of oxidation in the blood would be hindered by the circulatory and respiratory disturbances, the existence of which seemed to be proved by the results of autopsies ; he laid special stress on the presence of a double circulation through the open state of the foramen ovale, ductus arteriosus, and ductus venosus ; and he thought that the disease was very closely allied to icterus neonatorum. Valleix (221), also, regarded sclerema as a general malady consisting in disturbances in the circulatory and respiratory functions, and recognised as causes the action of cold and the presence of congenital debility. West (226) at first accepted this explanation, but in the seventh edition of his text-book (published in 1884) he expressed himself as follows : "I was once disposed to look upon the state of the lungs as furnishing the clue to all the phenomena of œdema of the cellular tissue. But the theory must, for several reasons, be given up as untenable, at least in the sense of its affording a constant and sufficient explanation of the causes of the affection. In the first place, changes in the pulmonary tissue are not constant, but are found only in about half the cases ; secondly, in many instances they

seem to be consequences rather than causes ; in the third place, while the changes are distinctly not pneumonic, they are far from being constantly those of mere collapse, but are at least as often those of intense congestion, sometimes even of pulmonary apoplexy." West leaves the problem unsolved, after remarking upon the extraordinary fall in temperature met with in sclerema, a lowering of the body-heat even greater than occurs in cholera. Depaul (122) regards the pathogenesis as complex, and seems to think that the essential factor is engorgement of the heart and vessels from weak cardiac and pulmonary action in a premature and congenitally feeble infant.

7. The Hepatic Theory.

Only a few words need be said with regard to the theories that explain the origin of sclerema by a reference to the state of the digestive system. Many of the older authors were struck by the association together of icterus and sclerema, and some—*e.g.*, Henke (189), Palletta (38), etc.—thought that the latter state was due to disordered hepatic functions. Léger (37) considered that jaundice of the new-born was a mild form of induration of the cellular tissue, and based his belief upon some experiments by Chevreul ; he also ascribed great etiological importance to shortness of the intestines ; but Billard and others long ago demonstrated the erroneous character of these theories. I need say nothing with regard to Parrot's explanation of sclerema (206), for that distinguished author was describing the hardening of the subcutaneous tissue which follows athrepsia and not true sclerema in the sense in which it is now generally understood. (See further under Diagnosis, p. 55.)

8. The Nervous Theory.

Although Underwood in his later writings (219) and Stütz (14) seem to have had some idea of a nervous theory in their views upon the nature of sclerema, Liberali (29) was the first to put this opinion definitely into words. The last-named writer thought that the action of cold paralysed the cutaneous nerve-tracts, so that the special vitality of the subcutaneous tissue was diminished, the fluids became dense, and, the freedom

of the peripheral circulation having been interfered with, there was congestion with all its results. J. F. Meckel (200A) regarded the morbid state of the skin in sclerema as only a symptom of imperfect vitality of the central parts of the nervous system. Vogel (223), whilst he considered the cutaneous changes as of the nature of an acute œdema due to general conditions of the new-born, also stated that as there was always, save in the cases complicated by pneumonia, diminished activity in the circulatory and respiratory systems, it was necessary to believe in a defect in the innervation of the cardiac muscle leading to slowing in the pulse-rate, with consequent chilling and exudation at the periphery. According to Steiner (217), sclerema is a general malady resulting from profound disorder of the respiratory and circulatory systems, originating perhaps in great part in a defect of innervation. Musmeci d'Agata (137) thought it might be regarded as a vasomotor and trophic neurosis of the vagus; and Angel Money (142) compared it to the myxœdemalike condition of skin met with in some cases of infantile paralysis, and in certain instances of exophthalmic goitre, and believed that it was produced by paralysis of the intercostal muscles and diaphragm, leading to interference with the respiratory functions. C. M. Campbell (166) regarded sclerema as a tropho-neurosis of the gravest character, possibly allied to that which in later years produces myxœdema; and I myself, writing in 1889 (147), thought that the primary pathological factor might be a trophic lesion of the nervous system.

It is now necessary to consider the theories advanced by the two Sommas. L. Somma (113) enunciated his views prior to the publication of those of Steiner, Money, and Musmeci d'Agata, but they were lost sight of till re-stated recently in a somewhat different form by G. Somma (150). L. Somma's explanation consists, indeed, of two theories,—a nervous and a bio-chemical. According to the former, sclerema is due to a vasomotor and trophic neurosis caused by morbid stimulation of the branches of the sympathetic, due to cutaneous chilling from atmospheric cold or the use of cold baths, and acting upon the delicate organic constitution of the new-born infant. In his second theory, L. Somma stated that cutaneous refrigeration led

to an inertia of the bio-chemical functions of the body which maintain the temperature of the organism, and so caused less production of heat, with a consequent lowering of the temperature.

G. Somma's theory is somewhat different. It is first pointed out that the body-heat is maintained by oxidation, regulated by the central nervous system. Some of the thermic centres (the excitatory) stimulate oxidation, and so increase the temperature; the others (the moderating) keep in check the processes of oxidation, and so tend to diminish thermogenesis. Somma then gives Baculo's observations on the localisation of the centres, and points out that lesions of the excitatory centres (probably located near the lateral ventricles and in the cortex) will produce hypothermogenesis and a fall in the body-temperature; whilst disturbance of the moderating centres (in optic thalami and anterior corpora quadrigemina) will lead to hyperthermogenesis. The normal temperature of the body is maintained when the activity of these two groups of centres is in equilibrium; if, however, the functional activity of the former be greater than that of the latter, there will be a fall in the body-heat, whilst if the latter predominate a rise will ensue. Somma then applies these facts to the elucidation of the pathogenesis of sclerema. He recognises two causes of the disease,—the one is predisposing, and consists in congenital debility of the infant; the other is determining,—it is cutaneous chilling from atmospheric and other causes. He states that the cutaneous chilling may be supposed to act in two different ways. According to the first theory, the abnormal stimulus excites the peripheral terminations of some of the centripetal nerves in the skin; these in turn transmit the effect to the excitatory thermic centres (lateral ventricles and cortex), and set up in these centres certain functional disturbances (either histological alterations or only molecular changes); then the stimulus is sent on by centrifugal nerves, and acts by reducing the processes of organic oxidation, the effect being most evident in the cellulo-adipose subcutaneous tissue stratum which lies nearest to the muscular layers. The second possible mode of action, according to G. Somma, is as follows: The cold producing cutaneous

chilling determines in the first place an ischaemia of the capillaries of the derma, whereby the blood flows to the internal organs, and thus in them all the circulatory disorders that have been described are set up ; in the second place, by the great weakening of the capillary circulation produced by the lowering of the temperature and by the congelation of the cutaneous adipose tissue, which will constitute an obstacle to the free peripheral circulation, there results the intense stasis along with all the conditions that the morbid anatomy and symptomatology of the disease have revealed.

Having in this way accounted for the oedematous variety of sclerema, G. Somma proceeds to apply his explanation to the indurative also. He refers to Langer's statement (123), which was to the effect that the fat of the new-born infant contains a greater proportion of the solid fatty acids (palmitic and stearic) than does that of the adult, and points out that it will require a higher temperature to keep the fat in a fluid condition in the former than in the latter, and that, therefore, a fall in the temperature which would be insufficient to cause solidification in the case of the adult may serve to do so in the new-born infant. Finally, he explains the preference shown by the morbid cutaneous changes for certain regions of the body (*e.g.*, the limbs, and especially the lower limbs) by pointing out that these parts are furthest from the heart, and that in them the body-heat will be dissipated sooner than in other regions.

In many ways G. Somma's theory commends itself as a reasonable explanation of the nature and origin of sclerema ; but it must, at the same time, be borne in mind that it is in some of its details hypothetical. Lesions in the thermic centres, for instance, have not been demonstrated ; and it is not certain even whether the localisation and mode of action of these centres is such as has been described. The occasional occurrence of truly congenital cases of sclerema constitutes, in my opinion, the greatest difficulty to accepting the above-described theory ; for G. Somma attributes great importance to the effect produced by cutaneous refrigeration, and it is very difficult, from what we know of intra-uterine physiology, to imagine such a chilling of the foetus as would be necessary to cause lesions of the thermic

centres, when it is borne in mind that the infant in utero truly lives in a warm water-bath. A very evident way to get rid of this difficulty would be to suppose that the disturbance of the thermic centres may occur before birth ; but G. Somma specially states that he does not believe in such a congenital disturbance of the nervous system, for the reason that in some cases several days may elapse after the birth of the infant before the scleremic phenomena (fall of temperature, etc.) appear. I think he is in error in denying this possibility, and in ascribing such a preponderating influence to cutaneous chilling. I believe that in most cases the morbid process resulting in sclerema is initiated in utero, that the most important factor in its pathogenesis is an antenatal lesion of the nervous system, possibly of the thermic centres, and that cutaneous chilling is only one of several conditions that may serve to develope the malady when it is not already present at birth.

Such are the many pathogenetic theories that have been advanced by writers on sclerema, and their number and variety serve well to demonstrate one thing at least,—the unsatisfactory state of our knowledge of the whole subject. It would seem as if nothing less than the labours of an international committee of investigation might succeed in clearing up the confusion, and in undoing the results of the erroneous generalisations of the past century.

CHAPTER IV.

SCLEREMA NEONATORUM—Continued.

DIAGNOSIS; PROGNOSIS; TREATMENT; LITERATURE.

DIAGNOSIS.

THERE ought to be little difficulty in recognising a case of sclerema neonatorum if the occasional occurrence of the malady is borne in mind, and if its distinctive clinical features have been clearly impressed upon the observer's mind. The association of marked lowering of the body-temperature, with depression of all the organic functions, and with the development of œdematos or indurative changes in the skin and subcutaneous tissue in an infant a few days old, points directly to the existence of this disease. Further, the diagnosis will be much easier if the observer have previously seen a case of the same nature. Since, however, sclerema has been constantly confounded with other pathological conditions, it will be well briefly to point out the chief factors in the differential diagnosis.

1. *From Erysipelas Neonatorum.*

There seems to be little doubt that many of the older writers confused together sclerema and erysipelas neonatorum ; thus the cases reported by Horn (16) seem to have been instances of the latter malady. Erysipelas, however, usually begins near the umbilicus or on the face, and rapidly involves the skin in the immediate neighbourhood ; the affected parts are red in colour, warm to the touch, and painful on pressure ; and the temperature of the body is raised. It must also be borne in mind that it is possible that erysipelas may occur as a complication of sclerema ; in such circumstances the diagnosis will of necessity be much more difficult.

2. From Scleroderma.

Whilst erysipelas is most likely to be confounded with the œdematous variety of sclerema, scleroderma resembles most closely the indurative type. The age of the patient will here be of the greatest diagnostic importance, for scleroderma rarely or never occurs before the end of the first year of life, while sclerema is met with almost constantly in the first month. Should, however, sclerema occur at a later period in life it could be distinguished from scleroderma by the following features :—The latter malady affects most commonly the upper half of the body, runs a comparatively slow course, has a not unfavourable prognosis, shows no important lowering of the body-temperature, and is not accompanied by marked depression of the organic functions. Further, the skin in scleroderma is of a wax-like colour, and rarely, if ever, is there any œdematous infiltration.

3. From Tetanus Neonatorum.

The resemblance between tetanus and sclerema is a superficial one. The body never becomes so stiff and immovable in the latter disease as in the former, and the use of the thermometer will clear away any doubts that may remain. At the same time, it is probable that some of the earlier reports of cases of sclerema included instances of tetanus.

4. From Induration of the Cellulo-adipose Tissue (so-called Sclerema Adiposum).

G. Somma (150) is of opinion that sclerema neonatorum ought to be differentiated from the induration of the subcutaneous and adipose tissue which occurs after death, or even occasionally in the ante-mortem agony in the case of infants who have suffered from exhausting maladies, especially of the nature of athrepsia or infantile diarrhoea. I agree with this writer as to the advisability of separating the two conditions ; but most authors, including such recent observers as Soltmann (215) and Max Runge (210), simply regard induration of the adipose tissue, Parrot's "endurcissement athrepsique" (206), and Soltmann's "symptomatic sclerema" (215), as a variety of sclerema. It is a

morbid process which occurs towards the close of a wasting malady, and not a disease which appears, as sclerema does, without prodromata and within a few days of birth. It is also truly a post-mortem phenomenon in some cases. It may be differentiated from sclerema by the time of its occurrence, as has been stated above, and also by the fact that it chiefly affects the parts of the body (front of the chest, back, upper and outer part of the limbs) that usually escape the scleremic change. The skin is white or yellow in colour. It cannot be pinched up in folds as it often can be in the œdematosus variety of sclerema ; but this fact is not of much diagnostic importance, for it is the indurative and not the œdematosus variety that is likely to be confounded with adipose induration. From these facts it ought to be easy to distinguish the disease from the cadaveric change if the patient has been under observation during life ; but it is very difficult for the medical jurist, for instance, to state definitely whether in the case of a dead body of an infant true sclerema has existed during life, or whether the conditions found are due to pathological changes occurring just before death (preagonalic) or just after it (cadaveric). G. Somma (150) gives the two following tests : (1) The preagonalic or cadaveric induration would disappear or diminish with the commencement of putrefaction ; and (2) it would also diminish on the application of heat to the dead body ; whilst if the infant had succumbed to true sclerema, early putrefaction and the application of heat would not so affect the corpse.

5. *From Cœdema Neonatorum.*

As I have already stated, there is a condition to which the name "œdema neonatorum" ought to be applied. That condition is not a disease *per se*, but simply a phenomenon met with in connexion with several maladies. It is œdema occurring in the new-born infant as one of the symptoms of cardiac, pulmonary, or renal disease, or of changes in the blood itself, or in the circulation in any special part of the body. It is certain that many of the cases which have been described as sclerema have really been of this nature ; but it is often impossible from the details given to eliminate the instances of œdema neonatorum

from the observations. The distinguishing character is to be looked for in the temperature. In sclerema the thermometer reveals a great, rapid, and progressive lowering of the body-temperature (external and internal), whilst in oedema there is not only not a fall in the body-heat but even in many cases a distinct rise above the normal. When there is a localised oedema due to some obstacle to the venous circulation, the condition may, according to G. Somma, be differentiated from the oedematous variety of sclerema by paying attention to the following points: in the former the skin may have a natural (rosy) colour, rarely reddish or bluish, it does not give to the touch the sensation of marble coldness, it keeps its elasticity, it takes again its natural outlines after having been compressed, it is painful on pressure, and the finger leaves an impression which only slowly disappears; in chronic cases, also, the skin may have cracked at various places, allowing a little serum to exude; and finally, a distinct morbid process may be found to account for the oedema. A comparison of the above-mentioned characters with those described in connexion with the oedematous variety of sclerema will in most cases enable a diagnosis to be made. There is an oedema neonatorum due to *garage*, but that is not likely to be mistaken for sclerema.

6. From General Dropsy of the Fœtus.

General fœtal dropsy is likely to be confounded only with the oedematous type of sclerema. The former malady, however, is always truly congenital, whilst the latter is only sometimes so. Further, general dropsy of the fœtus is much more evident and more deforming than is sclerema, and it is also accompanied by large fluid accumulations in the peritoneal, pleural, and pericardial sacs, which are absent or but slightly marked in the latter disease. If the fœtal anasarca be only slight, it may be very difficult to differentiate it from a congenital case of sclerema; in fact, it may be doubted whether it ought to be separated from that disease at all. *Congenital oedema neonatorum* is probably a form of general dropsy of the fœtus.

7. From Algidité Progressive (Hervieux).

Hervieux (83A) attempted to prove that the condition named

by him “algidité progressive” was a disease *sui generis* and quite distinct from sclerema. A careful examination of his paper, however, reveals the fact that whilst he emphasized the presence of a lowered temperature and a slowing of the heart’s action and of the respiration, he did not deny the existence of œdematosus and indurative cutaneous phenomena. It may be gathered, then, that his cases were really examples of sclerema as it is now understood.

PROGNOSIS.

Sclerema has been recognised as a very fatal disease by all writers ; and from what has been said with regard to its nature and causes, it is not difficult to understand that this should be so. It will be well to consider—(1), The mortality ; (2), the prognostic points ; (3), the duration ; and (4), the termination of the malady.

1. Mortality.

The percentage mortality in sclerema has always been very high, but it has varied considerably in different places and at different times. Soltmann (215) puts it, on an average, at from 80 to 90 per cent. ; and according to Casper,* out of 645 cases met with in the Hospice des Enfans trouvés in Paris between the years 1808 and 1811, 567 died. Matters had not improved in 1818, when out of 250 cases at the Parisian Foundling Asylum only 49 were saved. Out of 53 cases only 4 recovered—Elsässer, (81). The mortality was not, however, always so high as this ; thus, Andry stated that 9 out of 19 cases were saved, and Bruni, according to Loder (21), cured 55 per cent. in 1810 ; whilst Gölis (20), by means of antisyphilitic treatment, was able to report that not more than 8 out of 40 cases perished. In the second six months of 1823 Palletta (49), by the use of warm baths, etc., lost only 3 cases out of 62. In recent times Namias (133) saved 3 out of 7 cases. Robinson’s case (126) got well, and in the discussion which followed the reading of his paper, Weisse, Piffard, and Taylor also reported recoveries. It is probable that the average mortality is about 50 per cent.

* Casper (J. L.), *Charakteristik der französischen Medicin, etc.*, Leipzig, 1821.

2. Prognostic Indications.

The prognosis varies with the circumstances of the case, such as the constitution of the infant at birth, the variety of sclerema with which it is affected, the degree to which the temperature falls, etc.

(a.) An infant born at the full term with a healthy constitution will have a better chance of recovery than one that is weakly and premature.

(b.) The prognosis is better in the oedematous than in the indurative variety of sclerema, and with regard to the former there is more hope when the disease is simple and circumscribed than when it is complicated and widespread. The worst prognosis is arrived at when the infant is attacked with widespread sclerema of the indurative type, accompanied by complications. The congenital cases are more fatal than those in which the disease appears at a later date.

(c.) All writers are agreed as to the influence of the temperature : the lower it is the worse is the prognosis. Mignot (204) states that there is little hope if it fall 5° C. below the normal, and L. Somma (113) is of opinion that if it sinks below 30° C. recovery is impossible. G. Somma (150) believes that the gravity of the case is in direct relation to the lowness of the body-heat.

(d.) The presence of complications always increases the danger ; but the prognosis varies somewhat with the nature and severity of the concomitant diseases. According to G. Somma (150), lobular pneumonia, atelectasis, and fatty degeneration of the myocardium are more serious complications than *muguet*.

(e.) The better the climatic, social, and hygienic surroundings of the infant are, the better will be the prognosis. Thus, an infant born in the summer, fed in the natural way, properly clothed, and brought up in a private house, will have an enormously better chance of recovering from sclerema than one born in the cold months of the year, artificially fed, imperfectly clothed, and living in poverty or in a Foundling or Maternity Hospital.

3. Duration of the Malady,

True sclerema neonatorum always runs a very rapid course. Soltmann (215) gives its duration at from five to eight days, seldom longer. According to Andry (5), its average length is seven days. Exceptions, however, occur, and it may last for ten, fifteen, or even twenty days.

4. Termination.

As has been already stated, sclerema usually terminates fatally, but it may sometimes end in recovery. In the latter case the temperature begins slowly but continuously to rise, the circulation becomes more rapid, the respiration is easier, active movements of the limbs are more manifest, the cry is stronger, sucking and deglutition are more easily performed, and the skin begins to lose its intense coldness. The hardness of the integumentary structures in the indurative variety gets gradually less, and, according to G. Somma (150), passes through a stage of elastic hardness, and then through one of pasty density, till it reaches the normal. In the oedematous type the anasarca begins to disappear first from the eyelids and forearms, then from the thighs and hypogastrium, and then from the hands and legs, whilst it remains for a long time in the feet.

On the other hand, death may ensue ; then all the phenomena become more and more marked, and the fatal termination may occur quietly, or may be preceded by convulsions.

TREATMENT.

Since such widely different views have been held with regard to the nature of sclerema, it is not surprising to find that very many and various therapeutic means have been used to endeavour to combat the malady. Many of these have long since been abandoned, and at the present time the treatment consists almost entirely of attempts to counteract the fall in the temperature of the body which is so characteristic of sclerema. The remedial measures may be described under the headings of (1) the dietetic, (2) the hygienic, and (3) the medicinal (external and internal) means.

1. Diet.

In every case of sclerema the first requisite for the infant is a sufficient supply of healthy food. The mother's milk is, of course, to be preferred when it can be had, and the infant should be put to the breast every hour; where through weakness it is unable to suck, the milk should be drawn off and given in a spoon. If maternal feeding be impossible, as is in most instances the case, then an attempt should be made to secure the services of a good wet-nurse. Failing this, it is necessary to fall back upon artificial feeding with ass's or cow's milk; and it will be found most beneficial to sterilise it first. If the infant be so weak that it is unable to swallow the food, then an oesophageal or nasal tube must be employed, or recourse had to rectal feeding with peptonised milk.

2. Hygiene.

The environment of the scleremic infant ought to be made as satisfactory as possible. The patient ought to be placed in a dry, well-ventilated, and well-lighted room, the temperature of which ought not to be allowed to fall at any time below 22° C. The infant ought to be warmly clothed in flannel, or, what is preferable, should be wrapt up in cotton wool. Clementovsky (111) reports that in the Foundling Asylum in Moscow a sort of cradle with double metallic walls between which warm water is circulating is employed in the treatment of scleremic infants; and Andry (5), long ago, used to place them on a pillow near to an open fire. The best results, however, are to be expected from the use of an incubator, such as the *cuvette* of Tarnier.

*3. Remedial Medicinal Means.**(a.) External.*

For a long time the use of baths has been a popular means of treating sclerema. Roger (74) employed cold affusion; but nearly every one else has preferred to use warm baths. The temperature of the water ought to be from 32° to 35° C.; according to Eichhorst (178) it may be as high as 37° C.; the bath ought

to be repeated three times daily, and the infant ought to remain in it from fifteen to twenty minutes. Care should be taken that the temperature of the water is kept constant ; and G. Somma (150) recommends that a cold wet cloth should be held to the infant's head. The bath may contain simple water, or various ingredients may be added ; aromatic baths were frequently employed by the older authors—Carus (25), Breschet, etc.—and even at the present time mustard is sometimes put into the water. It is doubtful whether aromatic baths have any greater effect than those containing water only. Vapour baths were highly recommended by Underwood (219), Dugès (176A), Baron, Bouchut (164), and Roger (74) ; but Valleix (221) and others did not find that they produced greater therapeutic effects than the simple baths.

Sometimes warm lotions have been used in the treatment of sclerema ; thus, Clertan (82) employed one containing quinine ; and other writers have recommended the addition of salvia, lavender, marjoram, etc., to the fluid. The lotion was applied night and morning—Andry (5). Some—Capuron (167), Depaul (122), etc.—have spoken favourably of the use of fumigation, especially of the affected parts, with vapour of benzoin, gum ammoniac in acetic acid, etc.

Friction has been applied to the body in various ways. L. Somma (113) used warm flannel cloths and brushes, and Depaul (122) recommended the rubbing of the skin with alcohol or arnica. Roger (74) and Bouchut (164) advised friction with ice,—a somewhat heroic mode of treatment. The friction ought to be carried out in a special way, from below upwards, from the periphery to the centre.

Inunction has also been tried, and seems to have been beneficial in certain cases. Warm olive oil was used for this purpose by Marcacci (107), L. Somma (113), and Riedel (105) ; others used camphorated oil, or camphor ointment—Clertan (82), or liquor anodynus—Moscati (9). A few writers have spoken very highly of mercurial inunction, amongst whom may be mentioned Valleix (221), Gölis (20), and Carminati (35) ; the mercurial ointment should be rubbed twice daily into the skin of the legs, abdomen and thorax, and the application followed by a warm bath.

In yet other ways it has been attempted to raise the temperature of the skin and stimulate its functions, viz., by the application of hot bottles or warm sand-bags to the limbs of the child,—Roger (74), L. Somma (113), and others; by the use of counter-irritants, such as blisters—Girtanner (186)—applied specially to the indurated areas of the integument,—Andry (5), and Auvity (6); or by the employment of leeches placed on the thorax,—Billard (163), Palletta (38), Troccon (22), Léger (37), Dugès (176A), and others; or on the epigastrium—Denis (40), or on the mastoid process—Valleix (221). Multiple punctures of the skin have been advised, to allow the escape of the serum contained in the meshwork of the subcutaneous tissue—Ellis (179), but this procedure does not seem to be at all in keeping with modern ideas of the nature of the malady.

Perhaps the most successful methods of restoring the vitality of the subcutaneous tissue in cases of sclerema have been massage and passive movements of the limbs. Legroux (84 and 87A), Rochoux (209A), Namias (133), Soltmann (215), Demme (125), and many others have employed massage with good effect; and Galligo (184) and Descroizilles (173) have obtained satisfactory results from alternately flexing and extending the limbs for periods of from five to ten minutes, repeated every two or three hours. The faradisation of the paralysed muscles has been advised by Steiner (217), Eichhorst (178), and others.

Artificial respiration has also been recommended by Galligo (184) and Alberti (129), for the purpose of expanding the lungs, which have so often been found partially atelectatic.

Such are the external remedial measures which have been employed by various physicians and at various times, and it will be seen that their object is twofold,—first, the restoration of heat to the body, and, second, the re-establishment of the cutaneous circulation. Whilst warm baths, inunction, friction, and counter-irritants may all be employed, it will probably be found that the best results are to be obtained from the use of the conveuse and of systematic massage.

(b.) *Internal.*

The older physicians used to give to scleremic infants various

medicines by the mouth. Girtanner (186) recommended emetics such as ipecacuan; Hulme (8) used aperients such as rhubarb and magnesia; and diaphoretics and diuretics, such as liquor ammoniæ acetatis, spiritus etheris nitrosi, and tincture of digitalis, have been advised by Carus (25) and others. Underwood (219) long ago suggested that all medicines should be made warm by the addition of compound spirits of ammonia. Mercury, in the form of calomel, was given by Gölis (20), Henke (189), and Siebold (42); and cinchona was recommended by Jahn (194). The most useful medicines that can be given by the mouth are such stimulants as wine—Riedel (105) and others; brandy—Steiner (217), Vogel (223), and others; and ether—Galligo (184); these are to be administered in doses of one or two drops in a teaspoonful of milk.

Since in many cases scleremic infants are unable to swallow medicines, hypodermic injections have been recommended and practised by Eichhorst (178) and G. Somma (150). In this way camphor dissolved in oil of sweet almonds has been introduced into the system, and G. Somma gave injections of citrate of caffeine and ether alternately. The solution of caffeine was made by dissolving half a gramme of the citrate in ten grammes of distilled water. The solution was kept slightly warm, and a few drops were injected into the subcutaneous tissue. The last-named writer also states that Voisin recommended curara for the purpose of relaxing the vasomotor nerves, dilating the bloodvessels, and so raising the body-heat.

The bronchial mucous tract has been also utilised in the treatment of sclerema, and Bonnaire (149A) speaks highly of the curative effects of the inhalation of oxygen in this and in some other neonatal diseases. It may be concluded that this remedy acts beneficially by preventing the accumulation of carbonic acid in the blood and the consequent narcosis.

Probably the most useful medicines that can be given either by the mouth or hypodermically are stimulants such as brandy and ether, whilst the inhalation of oxygen and the injection of caffeine deserve a fair trial. It must be admitted, however, that the treatment of sclerema is still in a very unsatisfactory state.

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CHAPTER V.

DISEASES OF THE SUBCUTANEOUS TISSUE—
Concluded.

SUBCUTANEOUS ABSCESS IN THE FœTUS; DEFINITION, HISTORICAL NOTE, OLLIVIER'S CASE NATURE AND MODE OF ORIGIN: ATROPHY OF THE SUBCUTANEOUS TISSUE; DEFINITION, ILLUSTRATIVE CASE, NATURE AND MODE OF ORIGIN: "LIVING SKELETONS," ILLUSTRATIVE CASES: DERMATOLYSIS; DEFINITION, SYNONYMS, ILLUSTRATIVE CASES, "ELASTIC SKINNED MEN," CLINICAL FEATURES, PATHOLOGY, AND TREATMENT.

IN order to complete the consideration of the idiopathic diseases of the subcutaneous tissue in the fœtus it is necessary for me to discuss shortly two or three ill-defined pathological states, which are possibly curious rather than important. The morbid conditions referred to are: (1), Abscess in the subcutaneous tissue of the fœtus; (2), congenital absence or atrophy of the subdermal adipose layer; and (3), dermatolysis in the strict sense of the word. I might, also, have included here the other pathological conditions to which the name dermatolysis has been sometimes applied, and which have likewise been termed congenital elephantiasis, for many of them are essentially alterations in the subcutaneous tissue (of the nature of localised hypertrophies); but, as has been already stated (*v. Vol. I. p. 182*), these will be considered in the section of this work in which congenital tumours are dealt with.

SUBCUTANEOUS ABSCESS.

Definition.—Abscesses occasionally form in the subcutaneous tissue of the new-born infant in connexion with the progress of cephalhæmatomata, or as the result of traumatism during delivery; with these states we are not now concerned, but with the curious cases in which collections of pus have developed during fœtal life and have been found beneath the skin at the time of birth. Whether or not it is possible for such abscesses

to be formed apart from such general diseases as foetal erysipelas or intra-uterine tuberculosis is a question which it is difficult to answer, but concerning which something will be said immediately.

Historical Note.—C. W. Hufeland (*Journ. der prakt. Heilkunde*, lxiv., 1827) and H. F. Hardegg (*De Morbis Fœtus humani*, Diss., Tubingen, 1828) refer to the occurrence of furuncles or boils beneath the skin of the fœtus; and other writers, such as Ryan (*Lond. Med. and Surg. Journ.*, p. 485, 1834-5), Billard (*Des Maladies des Enfans*, p. 107, Brussels, 1835), Graetzer (*Die Krankheiten des Fœtus*, Breslau, 1837), and W. Raabe (*Die Krankheiten des Fœtus*, Diss., Marburg, p. 37, 1864) repeat this statement. These authors, however, give no details of individual cases, and it is to Ollivier that we are indebted for the account of an interesting example of what appears to have been a spontaneous subcutaneous abscess in the fœtus.

Ollivier's Case of Abscess in a Fœtus.—Ollivier and Serres * were asked to examine the body of a fœtus of about three and a half months, found in an enclosure near the Hôpital de la Pitié, in order to determine whether it showed any signs indicating that abortion had been criminally induced. The mother of the fœtus was unknown. Putrefaction was already somewhat advanced, but all the organs were quite recognisable, and none of them showed any appreciable morbid alteration. In front of the neck, on the right side, was a swelling of the size of a filbert, having a whitish-yellow colour, which formed a strong contrast with the rest of the surface of the body, which had a reddish tint. This swelling was soft and fluctuating, and extended from the base of the lower jaw to the right sterno-clavicular joint. An incision into it permitted the escape of a creamy pus, whitish in colour, and without odour. The pus lay immediately beneath the skin, separating it from the underlying muscular tissue. No alteration of the skin existed in front of or around the purulent focus.

Nature and Mode of Origin.—Ollivier was of opinion that the above-described condition did not justify the suspicion of

* Ollivier, *Archives générales de Médecine*, 2nd series, vol. v. p. 75, 1834.

violence, and thought that it could only have been the consequence of a phlegmon developed spontaneously in a foetus in utero. According to him it proved that in the early months of intra-uterine life the cellular tissue, although imperfectly formed, is already susceptible to inflammation, and may furnish all the elements for a purulent collection quite similar to those developed in later foetal life and after birth.

It is doubtful whether, in the absence of any history of the antecedents of the case, Ollivier was justified in forming these conclusions; at the same time it cannot be denied that inflammation may occur during intra-uterine life, as is proved by the occurrence of foetal peritonitis, of which I have myself seen a case.* It is, of course, taken for granted that Ollivier did not by any chance mistake the nature of the swelling on the neck, and that he was right in regarding it as an abscess, and not as, for example, a sebaceous cyst. Even if this be allowed, however, it is by no means certain that the collection of pus was not due to foetal erysipelas or tuberculosis. Certainly the statement that the collection of pus could only have been the consequence of a phlegmon developed *spontaneously* in a foetus in utero is open to criticism, especially when it is borne in mind that nothing was known of the mother or of the circumstances of the birth. Nevertheless the observation is interesting and possibly unique.

ATROPHIC STATE OF THE SUBCUTANEOUS TISSUE.

Definition.—Atrophic foetuses are not uncommonly met with as the result of disease of the placenta, of abnormal conditions of the umbilical cord (torsion, knots, etc.), and of morbid states affecting the mother during pregnancy; but in them the atrophy is usually general, and shows itself by the small size of the whole foetus and of its several organs. In the pathological condition now under review the atrophy affects only (or principally) the subcutaneous layer of adipose tissue, the size of the foetus being practically normal for the age of intra-uterine life arrived at.

Illustrative Case.—Instances of this morbid state are probably not uncommon, although doubtless they are seldom regarded as

* Ballantyne (J. W.), *Edinburgh Obstet. Soc. Trans.*, xv. p. 56, 1890.

of sufficient importance to warrant recording. In a case of maternal renal disease which I saw in December 1891 in consultation with my friend Dr Robert Stewart, the patient, after a pregnancy complicated by albuminuria, dropsy, and convulsions, gave birth to a male foetus corresponding in size to the calculated age of the gestation (6 months). The foetus, which was certainly alive shortly before birth, had a peculiarly shrunken appearance, the skin was everywhere closely applied to the underlying bones and muscles, and in the region of the face a marked senile expression was thus produced, and on dissection the subcutaneous tissue was found to be practically absent. It closely resembled an infant rendered atrophic by gastro-enteritis. The viscera showed no pathological appearances, and the serous cavities contained no fluid. The mother was a primipara, and had suffered from nephritis since childhood.

Nature and Mode of Origin.—That the lesion is due to mal-nutrition seems beyond doubt, but why the atrophic change should be limited to the subcutaneous tissue is less easy to understand, unless it be supposed that the cause of the atrophy only came into action shortly before the birth of the foetus, and affected the part of the body most liable to feel the result of imperfect nourishment. Wasting diseases affecting the pregnant woman may lead to an atrophic state of the subdermal adipose tissue in her infant; but it is at the same time a matter of frequent occurrence that a patient in the last stages of pulmonary phthisis, or *in extremis* from the incoercible vomiting of pregnancy, may yet give birth to a plump, well-nourished child. Probably the state of the placenta rather than the general health of the mother is the chief etiological factor in the production of the disease. As a matter of fact the placenta is often affected in chronic renal disease of the mother, and in the case above narrated the placenta was small and had a shrunken appearance. Unfortunately it was not examined microscopically.

“LIVING SKELETONS.”

A few words may here be said with regard to the individuals occasionally exhibited at Shows and Fairs, and known popularly

as “Living Skeletons,” for in some of them it is probable that we have to do with a congenital absence of the subcutaneous fat which has persisted till adult life.

Illustrative Cases.—In 1826, Claude Ambroise Seurat, the so-called “Anatomie Vivante,” or “Living Skeleton,” was exhibited in the Chinese Saloon, Pall Mall, London, and in other towns of the United Kingdom. He was then 26 years of age, about five feet and a half in height, and showed such a markedly atrophic state of the subcutaneous fat, and to some extent of the muscular system also, as fully to justify the designation “Living Skeleton” which was popularly applied to him. In one of the newspapers of the time the following statement occurs: *—“His father and his stepmother are both with him, and if we credit their account, he was brought into the world thus afflicted, grew to his present height at fourteen years old, and has never had a day’s illness in his life.” His parents, however, do not seem to have been very clear about the congenital character of the anomaly, for in another account, written for some paper by a medical man, the following sentence occurs: “According to the statement of his father (the mother is dead), he presented nothing extraordinary in his appearance at the time of birth; though, in my own mind, I have no doubt but the same malformation was existent then, which is so apparent now.” One of the advertisements with regard to this man reads thus:—

“ANATOMIE VIVANTE, OR LIVING SKELETON.—This extraordinary production of Nature has been examined by many of the most eminent of the Faculty in France and in England, who pronounce him a great phenomenon. He has been brought to this country at a considerable expence (*sic*), in the confident hope that he will contribute to the advancement of science. He is 28 years of age, in good health, and has from his birth resembled a Skeleton, although he has attained an enlargement of bone equal to any of his age.—The Faculty and the Public are invited to view so unparalleled a Being, who will be exhibited on

* For this and some other references of the same kind I am indebted to a collection of newspaper cuttings in the Library of the Royal College of Physicians, Edinburgh. The name of the collector is not known, and the book is entitled “Autographs.”

TUESDAY next, 9th inst., at the Chinese Saloon, 94 Pall Mall.—Hours of exhibition from 10 to 1, and from 2 to 6.—Admission, 2s. 6d."

Another "Living Skeleton" was Mr Calvin Edson, a native of the United States, who was to be seen in this country in 1830. Yet another was on show at Bartholomew Fair in 1828, "admission, one penny." With regard to these cases, and to many others which have been on view in "Shows," "Dime Museums," and at Fairs (often in association with "Fat Women"), nothing can be said as to a congenital origin, and probably many of them are artificially produced to pander to the popular taste for the *bizarre* and curious. At the same time, some of them may be due to a true ante-natal anomaly of the dermal and sub-dermal tissues.

DERMATOLYSIS.

Definition.—The term Dermatolysis was introduced by Alibert (*Monographie des Dermatoses*, 1832) to designate an affection characterised by an abnormal extensibility of the skin due to a particular alteration in the contractile faculty of that envelope; but the name has been employed in a wider sense to include the conditions also known as fibroma molluscum, molluscum fibrosum, molluscum pendulum, and congenital elephantiasis. Bazin (Article "Dermatolysie" in *Dict. encyclop. des Sciences médicales*, 1st ser., xxvii., p. 637, 1882) regarded Alibert's definition as incomplete, for the reason that it did not take into account the accompanying alteration in the dermis,—the increase, namely, in the number of certain of its anatomical elements; and he went on to assert that dermatolysis never takes place without a certain degree of hypertrophy. It is, however, preferable to separate dermatolysis from the hypertrophic states known as fibroma molluscum and congenital elephantiasis, for, as Crocker (*Diseases of the Skin*, 2nd edit., p. 587, 1893) points out, there are "congenital cases where there is loose attachment of the skin without hypertrophy, and it is to these that the term dermatolysis should be restricted." It is with these congenital cases of dermatolysis in this restricted sense that we have here to do.

Synonyms.—Leaving out of account the terms fibroma cutis, fibroma molluscum, etc.—which, although often regarded as synonymous with dermatolysis, really refer to different conditions altogether—it may be stated that the true synonyms are *cutis laxa* (Kopp and Seifert), *cutis pendula*, *cutis lapsus*, and *chalazodermie*. Individuals affected with this anomaly are popularly called “*Elastic skinned men.*” Meek’ren, who was probably the first to describe a case of this kind, entitled his communication “*De dilatabilitate extraordinariâ cutis.*”

Illustrative Cases.—The case reported by Meek’ren* in 1682 was that of a young Spaniard who could draw a fold of skin from his right breast up to his mouth and as far as his left ear, who could cover his face with a fold drawn up from the chin, and who could extend the skin over his right knee outwards for half a yard. The skin immediately returned to its place, and did not remain in folds. The affection was unilateral, for on the left side of the body the skin showed no such anomaly as on the right. A well-executed drawing representing the condition accompanied the letterpress. This individual was referred to also in Bell’s *Surgery* (vol. iii. p. 36, 1815), and in D. Turner’s *Treatise on Diseases incident to the Skin* (Introduction, p. x., London, 1736), and in other works of the last and of the present century. There is no statement regarding the congenital nature of the anomaly.

Another genuine case seems to have been that reported in the *Lancet* (vol. i. for 1882, p. 157, Jan. 28). The article, to which no author’s name is attached, is entitled “*The Elastic Skinned Man,*” and reads as follows:—“Mr Farini is now exhibiting at the Westminster Aquarium a middle-aged German. His appearance is in no way unusual, but he is able to draw the skin from any part of his body up in folds from one to five or six inches long. As examples of this may be given,—the skin of the forehead, which can be folded forwards for three inches; that over the nose, ears, the palm of the hand, and the back, can be drawn out into folds of varying length in the same way. The skin, too,

* Meek’ren (Jobi à), *Observationes medico-chirurgicæ* (Translation into Latin by A. Blasio), p. 134 Amstelodami, 1682.

is movable over the subjacent parts, so that on the limbs it can be glided up and down like the sleeve of a coat or leg of a trouser. There is no appearance of subcutaneous fat, and when one of these folds of skin is held in front of a light, the disposition of the cutaneous vessels, and also the absence of fat, can be plainly seen. . . . It is evident that the long folds of skin are not formed by the elastic stretching of the skin, for on observing one being drawn out, the skin from the whole surrounding part is seen to glide toward the fold and help in its formation. The skin is not specially elastic. The two situations where the condition is most inconvenient are the palm of the hand and the sole of the foot,—in the one interfering with firmness of grasp, in the other with security in standing."

Under the title of "Un cas de dermatolysis généralisée," W. Dubreuilh (*Annales de Derm. et de Syph.*, p. 529, 1887) described a case in which relaxation of the skin followed upon urticaria and œdema, but this was evidently an instance of the acquired, not of the congenital form of the disease. In the following year, however, Kopp (*Münchén. med. Woch.*, xxxv. p. 259, April 10th, 1888) gave a demonstration of two cases of "Cutis laxa," which seem to have been both congenital and hereditary. At any rate, the individuals affected were father and son; and the latter, who was eight years old, showed the anomaly in a less marked form than his parent. Kopp thought the condition might be due to an anomalous state of the elastic fibres of the skin, or to a congenital defect in the union between the skin and the underlying tissue (fascia and periosteum) affecting especially the strands of connective tissue.

Under the heading "Remarkable Abnormality in an Adult," J. A. Wetherell (*Brit. Med. Journ.*, vol. i. for 1889, p. 527, March 9, 1889, Illustrated) reported a case which Pye Smith (*Brit. Med. Journ.*, vol. i. for 1889, p. 743) regarded as dermatolysis, and as similar to the example seen by Meek'ren. I do not, however, feel certain that Wetherell's patient really exhibited dermatolysis in the strict sense of the term. He was a man of forty years of age, the right half of whose chest was obscured from sight by a large overhanging fold of tissue. He stated that it had existed from birth, that at first it was small, and that its

growth had been proportionate to the rest of the body. It caused him no inconvenience save that after work he felt a dragging sensation in it; and he generally wore a binder to keep it in place. With regard to the fold itself, it began at the right side of the sternum, its upper border ran in one continuous piece, with a margin of 4 inches of healthy tissue, from the clavicle along the thorax, then bending at an obtuse angle grooved the inner limit of the biceps for a distance of 10 inches—taking the acromion as the fixed point. The lower border was 18 inches from the upper, and reached the fold of the groin, where it was reduplicated and returned upwards to within one inch of its commencement. The entire circumference of the larger mass was 20 inches; that of the arm portion which skirted posteriorly the inner edge of the triceps was 10 inches. There were numerous folds and wrinkles. On raising up the greater bulk a normal nipple was discovered exactly at the middle line, 9 inches from its lowest point. The entire surface was dotted with comedo-like black points, occupying the situation of the hair follicles; it was, however, devoid of hair excepting a patch on the superior internal angle and the upper line of the arm-piece. The left axilla and hypogastric region were also hairless. The colour was a shade deeper than that of the surrounding skin; cold corrugated it, producing a condition like cutis anserina. Wetherell regarded it as a supplementary portion of a breast, which, somewhat misplaced, had gradually, during its intra-uterine existence, fallen by dint of its own inherent weight over the remainder. It was a unilateral excrescence, having had an intra-uterine life. With Wetherell's view I am inclined to agree rather than with Pye Smith's, and in all probability the case was not one of dermatolysis in the strict sense of the term, but of *fibroma molluscum seu pendulum*.

A genuine case of dermatolysis seems to have been that reported by Otto Seifert ("Uber cutis laxa," *Centralblatt für klin. Medicin*, xi. p. 49, 1890). The patient, a man of 19 years of age, showed over his whole body an abnormal folding and dilatability of the skin. The somewhat dry integument could be folded or pulled out, and then returned to the normal again when the extending force was removed. A piece of skin from

the anterior chest wall was removed and examined microscopically, when it was found that the elastic fibres were quite normal, whilst there was a transformation of the firm tissue of the dermis into an unformed tissue like a myxoma, along with the total disappearance of the connective tissue bundles.

Finally, at the Waverley Market in Edinburgh during the Christmas week of 1892, a young man was exhibited who showed in a marked form this curious anomaly. He was called "Pierre, der anatomische Wundermensch," and he could draw out large folds of skin from his face, neck, chest, and limbs. He did not otherwise present anything unusual in his appearance. The affection in him was bilateral and doubtless congenital.

Clinical Features.—An idea of the clinical features of this morbid condition will have been formed from the perusal of the illustrative cases given above. The chief feature is, of course, the extraordinary extensibility or dilatability of the skin. The integument may on this account become detached and fall down by its own weight; but it is probably more common in the genuine cases of dermatolysis to find displacement only when a drawing out force is in action. The skin does not lose its functional activity: its colour, temperature, and secretions usually remain normal. According to Bazin (*loc. cit.*) its sensibility is a little diminished. The same writer also states that it is always partial; but he is evidently speaking of the hypertrophic conditions to which the name dermatolysis is loosely applied, for in the man "Pierre" and in some others the extensibility of the integument affected the whole body. The inconvenience in grasping and locomotion caused by the abnormal state of the skin has been already referred to.

Pathology.—Alibert (*Monographie des Dermatoses*, 1832) was evidently undecided as to exact pathology of dermatolysis, for he placed it in the group of heteromorphous dermatoses along with ichthyosis, tylosis, warts, and nævi. The anonymous writer in the *Lancet* (vol. i. for 1882, p. 157) considered that the causes of the peculiarity were the absence of the usual adipose layer under the skin and the extreme looseness and length of

the fibrous processes fixing the skin to the deep fascia. The skin is not really more elastic, for although Kopp (*loc. cit.*) expected that changes would be found in the elastic fibres in it, Seifert (*loc. cit.*) was able to state that microscopic examination showed them to be normal. The latter writer discovered that along with complete atrophy of the connective tissue bundles of the skin there was a transformation of the firm tissue of the dermis into an unformed myxoma-like structure. The presence of this ill-formed gelatinous tissue is according to Crocker (*loc. cit.*) a strong argument in favour of the view that dermatolysis is closely allied to fibroma molluscum. There is, however, one great distinguishing feature, which is, that in the latter there is a hyperplasia of the connective tissue of the deeper layer of the corium and of the subcutaneous layer which is absent in the former. I am inclined to regard dermatolysis as consisting essentially of a weakening of the structural elements which normally bind the skin to the underlying structures, caused possibly by some fault in nutrition or circulatory disturbance (*œdema?*) occurring during foetal life. I have, at any rate, placed this condition on the boundary line between diseases of the subcutaneous tissue and those affecting the skin. I have also designedly considered it in the same chapter with congenital atrophy of the subcutaneous tissue. Something very similar to it is seen as an acquired condition in the abdominal walls and mammae of many multiparous women.

Treatment.—When the formation of hanging folds has not taken place no treatment is specially called for ; but when such folds are present, it may be considered necessary, both for the sake of appearance and comfort, to excise them and bring the parts together with sutures.

With the following chapter I commence the study of the idiopathic congenital diseases of the skin ; but something will be said concerning the development of the subcutaneous tissue and its bearing upon the above-described maladies at the close of this volume.

CHAPTER VI.

IDIOPATHIC DISEASES—Continued.

DISEASES OF THE SKIN.

ICHTHYOSIS : VARIETIES—FœTAL ICHTHYOSIS (GRAVE TYPE) : DEFINITION ; SYNONYMS ; HISTORICAL NOTE ; FREQUENCY ; CLINICAL HISTORY ; PATHOLOGY.

IT is convenient to begin the consideration of the diseases of the skin that may be met with at the time of birth with the study of the conditions that have been grouped together under the name of "congenital ichthyosis." During the last ten or twelve years great advances have been made in our knowledge of ichthyosis as it is met with in the fœtus and in the infant at birth : the connecting links between typical ichthyosis fœtalis and ichthyosis simplex of the adult have been recognised, and it has been almost conclusively demonstrated that the former affection is the same in its pathology as the latter. I shall consider, first, the typical grave form of foetal ichthyosis which gives to the infant so affected a distinctly monstrous appearance, and which almost invariably proves fatal within a few days after birth ; second, the less marked variety, the so-called attenuated form, which is often quite compatible with extra-uterine life, and which forms the connecting link between the first variety and ichthyosis simplex of the adult ; third, ichthyosis hystrix, traces of which are occasionally visible at birth ; fourth, the condition which has sometimes been called the minor degree of ichthyosis hystrix, but more often nerve-nævus or neuro-pathic papilloma ; and, fifth, keratosis pilaris.

ICHTHYOSIS FŒTALIS (GRAVE TYPE).

Definition.—The grave form of foetal ichthyosis may be provisionally defined as a skin disease of the fœtus, developed probably about the fourth month of intra-uterine life, characterised by the existence over the whole surface of the body of horny epidermic plates, separated from each other by fissures and

furrows, associated with certain deformities of the mouth, nose, eyes, ears, and extremities, and leading to the death of the infant very soon after birth.

Synonyms.—The first writers to describe specimens of foetal ichthyosis did not give to them this name, but employed much more general designations. Richter (1) regarded the morbid state as a *deformity* or *monstrosity*, Steinhausen (3) entitled his dissertation “*De Singulari Epidermidis Deformitate*,” and G. Vrolik (5) described his specimen as “*a singular defect in the skin*” (“een zonderling gebrek in de Huid”). Hinze (2), however, stated his belief that the affection was a *lepra* (*Lepra Græcorum*, or *Elephantiasis leprodes*), an opinion adopted by Feiler* in 1814; and Behrend (4), writing in 1839, gave to it the special name of “*cutis testacea*.” Graetzer (42), whilst he placed the cases of Hinze, Richter, and Steinhausen together under the designation *Elephantiasis*, also pointed out the resemblance which existed between those cases and the ichthyotic subjects known as “*Porcupine Men*.” Sievruk (8) named the disease a congenital hypertrophy of the epidermis (“*Hypertrophia epidermidis congenita*”). Keiller (7) simply called his case one of “thickening and deep fissures of the skin in an infant at birth;” but J. Y. Simpson (7), who communicated Keiller’s case, entitled his paper “*Intra-uterine Cutaneous Diseases*,” and went on to say that “it would appear to be much more analogous to Ichthyosis than to any other skin disease that can be referred to,” and therefore suggested for it the name of “*Ichthyosis Intra-uterina*.” This designation has been adopted by Smellie (11), Smith (26), Thibierge (49), and others; but the synonym “*Ichthyosis congenita*” has been more generally employed,—Müller (12), Houel (13), Schabel (16), Jahn (17), Löcherer (22), Stühlinger (28), Straube (31), Lassar (33), Bruck (34), Schab (35), Oestreicher (37), and Carbone (40). Unfortunately, *congenital* ichthyosis is the name which has been given in a loose sense to ichthyosis as it occurs in later life, on account of the fact that there is usually a congenital *predisposition* to it. F. Hebra (45) did not believe that the malady was ichthyosis, but regarded it as “*general*

* Feiler (J.), *Pädiatrik*, p. 23, 1814.

seborrhœa," and his belief and nomenclature have been recently adopted by Bland Sutton (32). Wilks (51), Nayler (18), Sutton (32), and others have also used the singularly descriptive name "Harlequin Fœtus." Lebert (44), having regard to the nature of the morbid process, suggested the term *Keratosis*, and called the affection "*Keratosis diffusa epidermica intra-uterina*;" Kyber (25), regarding the morbid production as a neoplasm, used the word "*Keratoma*" instead of Keratosis ("das universale congenitale Keratom der menschlichen Haut"), and both Wheelock (29) and Livingstone (30) have employed Kyber's nomenclature ("*diffuse congenital keratoma*"). Bar (41) and others* have preferred the expression "*fœtal ichthyosis*," and I have adopted this name on account of its brevity, and, as I believe, correctness.

Historical Note.—In enumerating the names that have been given to foetal ichthyosis I have already indicated something of the history of the disease. The first specimen seems to have been that put on record in 1792 by Richter (1) in his "*Disseratio de Infanticio*," but the description given of it was somewhat imperfect and unsatisfactory. Hinze (2) gave a much more complete account of his case. Steinhausen (3) in 1828, and Behrend (4) in 1839, each recorded an example of this disease. The microscopic characters of Steinhausen's specimen (preserved in Anatomical Museum of Berlin) were worked out some years later by Simon. Between the years 1840 and 1850 appeared a series of cases reported by Vrolik (5), Souty (6), Keiller (7), J. Y. Simpson (10), Sievruck (8), and Smellie (11); and during this decade a considerable advance was made in our knowledge of the disease, chiefly through the excellent monograph of Simpson. H. Müller (12) described a specimen with considerable fulness in 1850; Houel (13) recorded a case at Paris in 1853; Okel (14) noted two examples of the disease in the same family in 1854; and Schabel (16), in a dissertation of Stuttgart, gave a fairly complete account of the naked-eye and microscopic appearances of his specimen, illustrated by a good plate. Lebert (44), although he added no new case, rendered a

* Barkow (17A) called his case "*Ichthyosis fœtalis scutata pemphigaea*."

useful service by gathering together nine of the previously reported examples, and by drawing from their consideration certain general conclusions as to the character of the disease ; he also referred to instances in which this affection had appeared in the foetal calf. In 1869 Jahn (17) gave, in the form of an inaugural dissertation of Leipzig, a very complete account of a case of foetal ichthyosis, dwelling specially upon the microscopic appearances of the skin.

Between 1870 and 1880 little of note was written on the disease. In 1871 Barkow (17A) described a case in which there was also pemphigus. Nayler (18) referred to four specimens in Guy's Hospital in London ; Löcherer (22) shortly described a case in 1876 ; in the same year F. Hebra (45), in his text-book, stated his belief that this malady was not ichthyosis, but general seborrhœa, an opinion to which the author's great reputation as a dermatologist added much weight ; F. Hebra (23) also figured in his *Atlas* a not very typical specimen which had occurred in C. Braun's Clinique ; Houel, in combination with Chambard (24), described another example in 1878 ; and in 1879 Pelletier (46) discussed the disease in a thesis of Paris.

Kyber's communication (25), which appeared in 1880, was a most valuable one, but was sadly deficient in references to the work of earlier writers. Stühlinger (28) in 1880, Straube (31) in 1883, Bruck (34) in 1888, and Schab (36) in 1889, all published cases of foetal ichthyosis in the form of inaugural dissertations. Smith (26), Wheelock (29), and Livingstone (30) were the first writers to describe specimens of this foetal affection in America, and of these Livingstone's monograph was especially complete and thorough. H. Hebra (47) in 1883 published a paper on the subject, in which he quoted largely from Kyber's work, and returned again to the consideration of the disease in his *Lehrbuch der Haut—und Geschlechtskrankheiten*, which appeared in 1884. Bland Sutton (32) showed a specimen to the Medico-Chirurgical Society of London in 1886, and in the same year Lassar (33) gave a "Demonstration eines Präparates von sog. Ichthyosis congenita" to the *Berliner Medicinische Gesellschaft*. Thibierge (49) in 1889 published in the *Dictionnaire Encyclopédique des Sciences médicales* what is perhaps the

best monograph which has yet appeared upon "Ichthyose intra-uterine," and in the same year a good account of the London specimens was given in the third edition of Wilks and Moxon's *Lectures on Pathological Anatomy* (51).

Since 1890 a few cases have been described : one by Carbone (40) in Italy in 1891 ; in the same year three by Oestreicher (37, 38, 39) ; and one by Bar (41) at Paris in 1892.

Frequency.—It would seem that the grave form of foetal ichthyosis is a rare disorder, for I have been able to find no more than forty-two reported cases. No specimen has been shown in Edinburgh since that obtained by Dr Keiller in 1843. It must, I think, be granted that when such cases occur they are almost certain to be put on record, for the appearances are so striking that they cannot be overlooked, and so characteristic that they cannot be mistaken for anything else. So that there is little doubt that specimens of foetal ichthyosis are really very rarely met with.

CLINICAL HISTORY.

In discussing the symptomatology of foetal ichthyosis it will be convenient to consider, first, the maternal history ; second, the paternal ; third, the family history ; and fourth, the symptoms manifested by the infant himself. Some of the reported cases consist solely of the description of the specimen, which is then usually an old museum preparation, the clinical history of which has long been forgotten. This remark applies to the London specimens noted by Nayler, Wilks, Simpson, and others (18, 19, 20, 21), and to those figured or described elsewhere by F. Hebra (23), Straube (31), Schab (35, 36), and Bar (41). With regard to the remaining cases some details of the clinical history are forthcoming, although these are often very imperfect.

A. *Maternal History.*—The age attained by the mother when she gave birth to an ichthyotic foetus was occasionally noted : it was 24 in No. 30 ; 26 in No. 5 ; 34 in Nos. 2 and 40 ; 35 in No. 17 ; and 36 in No. 22. In most instances her *general health* prior to the pregnancy which terminated in the birth of the diseased foetus was good (Nos. 2, 3, 5, 6, 7, 10, 11, 14, 22, 25, 28,

30, 32, 33, 34, and 40), and in some cases it was specially mentioned that her skin had always been free from eruptions (Nos. 2, 10, 22, 30, 34, and 40); but in No. 17 she had suffered from rickets when a child, and in Nos. 2 and 26 there was a suspicion of the syphilitic taint. In No. 26, also, the mother, a blonde, was of a very nervous temperament. In no case had the mother suffered from a skin affection similar to that in the foetus.

With regard to the *sexual history* of the mother little information is usually forthcoming. In No. 17 menstruation began at the age of 16 and was always irregular, stopping for periods of from two to three months at a time. The previous pregnancies were occasionally referred to,—thus the mother was a i.-para in Nos. 5, 25, 30, and 40; a ii.-para in No. 14; a iii.-para in Nos. 6, 15, 17, 26, and 28; a iv.-para in Nos. 2, 7, 22, and 37; a v.-para in No. 38; a vi.-para in No. 39; an viii.-para in Nos. 33 and 34; and a “multipara” in Nos. 10 and 32. In most cases the previous pregnancies were normal, but in No. 7 the second gestation ended prematurely in the birth of a dead infant, and in No. 17 the first pregnancy had a similar termination (ascribed to the lifting of a heavy weight), and in Nos. 33 and 34 there were six previous normal gestations and one abortion. Whilst, therefore, it is true that the early obstetric history was in the great majority of instances good, one striking fact remains to be noted in this connexion. This fact was the occurrence of more than one ichthyotic infant in the same family. Thus Okel's two specimens (14, 15) were borne by the same mother (who had previously had one well-formed infant) within two years; Houel's two cases (24) were also the offspring of one mother; and, finally, the mother in Oestreicher's cases (37, 38, 39) had three normal infants by her husband, but after his death she gave birth to three ichthyotic foetuses in three successive years; these were illegitimate, but were presumably the offspring of the same father. These examples of “family-prevalence” are very interesting.

Fœtal ichthyosis does not appear often to have interfered with the progress of labour. The condition of the head, it is true, sometimes rendered the diagnosis of the presentation a difficult matter, as in Nos. 2, 7, 26, 30, etc., and the labour itself was

occasionally prolonged by the unyielding character of the foetal integument (Nos. 1, 2, 26, etc.). The child was, however, always born by the natural efforts alone, with the single exception of No. 30, in which instruments were required. The presentation was probably almost always cephalic. The pregnancy which terminated in the birth of an ichthyotic foetus was more frequently rendered abnormal than was the labour. It very often ended prematurely : at the 7th month in No. 7 ; between the 7th and 8th months in Nos. 13, 25, and 28 ; at the 8th month in Nos. 1, 5, 11, 15, 17, and 40 ; "prematurely" in Nos. 8 and 9 ; and at or near the full term in Nos. 2, 3, 10, 14, 17A, 22, 26, 30, 32, and 34. It is a striking fact that no foetus suffering from ichthyosis has been born at an earlier date than the seventh month of intra-uterine life, although it is almost certain that the morbid process itself begins about the fourth month. Besides its premature termination, the pregnancy was occasionally morbid in other ways : thus, in No. 1 the mother had suffered from sharp pain in the right side of the abdomen ; in No. 6 she received a severe fright at the fourth month, followed by intense abdominal pain, great disgust for food, and constant feeling of suffocation, along with a yellowish foetid vaginal discharge ; in No. 7 she was seized with acute pains in the left iliac region about the fourth month of pregnancy, and had a feeling that "something very unusual was the matter ;" and in No. 17 she noted the presence of very violent foetal movements, and thought that she was pregnant with twins. With regard to the occurrence of maternal impressions, it was noted by Nayler in connexion with No. 18 that the mother had been frightened at a country fair at the time of quickening ; in No. 26, when about two months pregnant she was much alarmed by stepping in the dark upon a poodle, and had thereafter a firm belief that the infant would be deformed,—her abdomen at the fourth month was much larger than normal ; and in No. 28 the mother was terrified at an early stage of gestation by the sight of a tortoise. It was definitely stated that there was hydramnios in Nos. 17 and 26 ; but possibly this occurred in other cases also.

It may be concluded that in most cases the puerperium was normal, although the fact was definitely stated only in Nos. 2, 7,

and 17; but in one instance (No. 30) the mother died on the seventh day post-partum, probably from septic peritonitis (the labour had been much prolonged on account of the want of lubrication of the passages, instruments had been employed, and rupture of the cervix and perineum had occurred). The later obstetric history was mentioned in only one case (No. 7), in which the mother again became pregnant, and was delivered of a normal infant in a normal manner.

B. Paternal History.—The father's health was good in Nos. 5, 6, 7, 10, 11, 14, 25, 28, 30, and 40. In No. 2 it was stated that the husband of the woman who gave birth to the ichthyotic foetus was a confirmed drunkard; but this statement was of no clinical value, for it was said further that he was not the father of the infant. In No. 26 the father had suffered from a soft chancre. It is probable that in all the other cases the father was healthy, at any rate as regards the state of his skin. The father and mother were blood relations (uncle and niece) in No. 40; and it is noteworthy that in No. 37 the mother had three healthy infants by her first husband and three ichthyotic foetuses by her second.

C. Family History.—The interesting fact of the occasional occurrence of foetal ichthyosis in two or more children born to the same parents has been already noted, and there is little else in connexion with the family history that merits attention. In No. 26 the mother's eldest infant had a nævus, but this was probably nothing more than a coincidence. In no case is there the report of the existence of ichthyosis or other skin disease amongst the parents' relatives, and in Nos. 11, 22, 26, and 34, the absence of such morbid tendencies is specially mentioned. The consanguinity of the parents in No. 40 has been already referred to.

D. Infantile History.—It is very remarkable that there is in only one instance (17A) the record of the birth of a dead or asphyxiated ichthyotic foetus,—in every other case the infant seems to have been born alive, and to have lived for some days, or at least hours; this is the more strange when it is borne in mind that premature labour was a very common occurrence. Ichthyosis is evidently not a cause of intra-uterine death. On the other hand, no infant

affected with this grave form of ichthyosis long survives its birth. The length of time in the recorded cases varied from nine days to a few hours : it was 9 days in No. 17 ; 8 days in No. 15 ; 7 days in No. 11 ; 4 days in Nos. 2 and 12 ; $3\frac{1}{2}$ days in No. 3 ; 3 days in Nos. 1, 26, 33, 34, 37, and 40 ; a "few days" in Nos. 8, 9, and 10 ; 2 days in Nos. 6 and 22 ; 36 hours in No. 25 ; 20 hours in No. 28 ; 12 hours in No. 7 ; 10 hours in No. 24 ; 6 hours in Nos. 14 and 30 ; and 3 hours in No. 5.

The *sex* of the ichthyotic foetus was not always mentioned ; but of the twenty-two cases in which it was noted, in eleven (Nos. 5, 7, 9, 11, 22, 24, 26, 28, 31, 36, and 40) it was male, and in the same number (Nos. 1, 2, 3, 8, 12, 16, 17, 17A, 25, 30, and 35) it was female.

In some cases (*e.g.*, Nos. 2, 34, 36, and 40) the infant was weakly at birth ; but in others it gave signs of considerable vitality, as shown by the activity of its movements. The child usually cried loudly and nearly continuously (Nos. 7, 10, 11, 14, 15, 17, etc.) ; but in some instances the cry was feeble and whining (Nos. 6, 30, and 40), and in others it was scarcely human, being described as a buzzing sound in No. 5, and as resembling the bark of a dog in No. 26. Respiration was generally impeded on account of the blocking of the nostrils with epidermic masses ; but it was described as easy in No. 6. The power of suction was usually absent, so that the child could not take the breast, but had to be fed with a spoon ; but in No. 11, whilst it was unable to suckle at first, it could do so later, probably on account of the separation of the epidermic plates, which had surrounded and prevented the opening of the mouth. In nearly every case, however, the infant, whilst unable to suck, could swallow easily (*e.g.*, Nos. 2, 6, 10, 11, 17, 22, 26, etc.), and took with avidity the fluids placed in its mouth. Exceptions to this rule were found in No. 1, in which deglutition was impossible, and in Nos. 14 and 40, in which it was feebly performed, and with difficulty. When the ichthyotic infant lived long enough, it usually urinated and defaecated naturally, and the urine was clear and odourless (No. 2), whilst the motions consisted of the usual green meconium. In Nos. 6 and 14, however, no urine was passed. In No. 30 meconium was vomited as

well as passed from the bowels. In most cases the infant slept little, and in some instances (1, 6, 17, etc.) special reference was made to the highly offensive, cadaveric smell which came from its skin.

Such were the symptoms associated with foetal ichthyosis (the external appearances will be described under the head of "Pathology"), and in none of the recorded cases was there anything like a complete clinical history. Jahn (17) gives what is perhaps the least imperfect account of the phenomena which occurred during the brief life of the infant; but a complete and carefully recorded statement of the symptoms of foetal ichthyosis is still a desideratum.

PATHOLOGY.

In most of the recorded cases of foetal ichthyosis the pathological appearances of the specimen are very fully given; perhaps the best descriptions are those found in Nos. 2, 3, 5, 6, 7, 12, 16, 17, 25, 30, 35, and 40. Thibierge (49), also, although he described no new case, gave an exhaustive account of the morbid anatomy of the disease in his monograph.

A. MORBID ANATOMY OF THE FOETUS.

I. *Macroscopic Characters.*

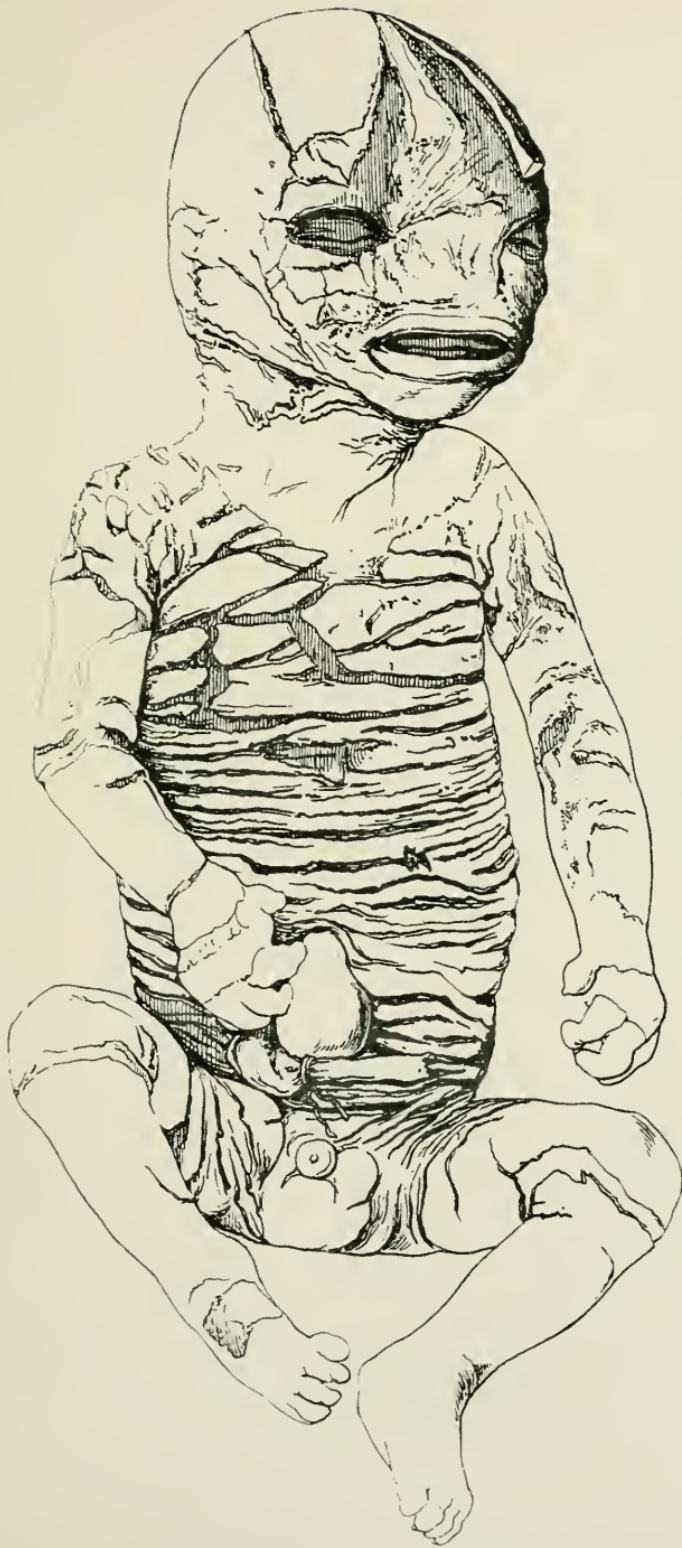
1. *Dimensions and Weight.*—Some writers have described the ichthyotic infants seen by them as of small size and puny, but others have noted that they were of average size and weight (24, 32). The exact weight was given in some instances: it was 9 lbs. in No. 16; 8 lbs. 6 oz. in No. 2; 3564 grammes in No. 30; 2650 grms. (without the viscera) in No. 34; 2655 grms. in No. 6; 4 lbs. 14 oz. in No. 17; 1625 grms. in No. 31; 3½ lbs. in No. 12; 1265 grms. (?) in No. 28; and 1 lb. 8 oz. (?) in No. 3. With regard to the total length of the infant, it was 68·5 cms. (?) in No. 2; 50 cms. in No. 34; 47 cms. in No. 16; 45·5 cms. in No. 3; 44 cms. in No. 17A; 43 cms. in No. 31; 41·7 cms. in No. 28; 40·5 cms. in No. 12; 40 cms. in No. 17; 39·5 cms. in No. 40; 39 cms. in No. 6; and 36·5 cms. in No. 25. In one or

two cases detailed measurements of the head, etc., were given : in No. 30 the O.F. diameter was 11·5 cms., the O.M. 12·75 cms., and the Bi-P. 9 cms. ; the transverse diameter of the body at the shoulders was 13 cms., etc. ; and in No. 31 the maximum circumference of the head was 30 cms., the O.M. diameter 10·3 cms., the Bi-P. 7·8 cms., and the Bi-T. 6·7 cms. These numbers are little, if anything, below the average, for it must be remembered that most of the infants were premature ; and an analysis of the cases in which the size and weight were given shows that the largest and heaviest infants (*e.g.*, No. 2) were those that were born at or near the full term of pregnancy, whilst the smallest (*e.g.*, No. 25) were expelled prematurely at the seventh month.

2. *External Appearances.*—Since foetal ichthyosis is essentially a skin disease, it follows that its most characteristic features are recognisable on inspection. The external appearances will, therefore, be fully described, whilst the visceral lesions will receive less attention. Since, further, all the recorded specimens resemble each other very closely (so closely that some writers have pointed out that it would be easy to suspect the authors of plagiarism), it will only be necessary to give one general description of the appearances found, indicating the small points of differences exhibited by certain of the cases (*vide* Plates II. and III.).

An ichthyotic foetus or new-born infant presents a particularly hideous and repulsive appearance, so that it is no matter for wonder that the older writers regarded it as a monstrosity, and applied to it such descriptive terms as terrible (1) and horrible (2). The whole body, from head to foot, is covered with a thickened, hardened integument split up into plates and scales by deep fissures and shallow furrows ; and the eyes, nose, mouth, anus, feet, and hands are markedly deformed. It will be well to consider first the appearances of the integument which are common to the whole body, and thereafter to describe its special characteristics as seen in the various regions.

As has been said, the whole integument is greatly thickened ; but the hypertrophied epidermis is everywhere divided into plates of varying size and shape by more or less wide and deep



STRAUBE'S CASE OF FETAL ICHTHYOSIS.

furrows, cracks, and clefts. The appearance thus produced has been variously compared to that of the bark of some trees (12), the dermal covering of the armadillo (16), the coat of the testudo or tortoise (33), and the party-coloured dress of the harlequin. Any resemblance to a fish is very difficult to find, so that in this respect the name "Ichthyosis" is a misleading one. Some authors (3, 22, etc.) have been struck by its likeness to an ancient coat of mail.

The colour of the surface of the body has been described as white (26), whitish (25), yellow (1), yellowish (2), dirty yellow (30), yellowish-white (31), yellowish-grey (16, 17, and 34), dirty grey (28), and greyish-brown (12). No doubt these differences in the descriptive terms used are due to the slight changes in colour which occur after birth ; when the infant is born it is more or less white in colour, for observers have at first sight regarded the covering as one of vernix caseosa, but in a few hours or days a more or less marked yellow tint appears. The integument, however, is not universally grey or yellow, for the intersecting furrows and fissures have a colour varying from red to blue (1), from violet to purple (6), or from red to reddish-brown (31).

In consistence the epidermis is much harder than usual. It has entirely lost its elasticity. Authors have applied various descriptive terms to it, such as leather-like (2), horny (12, 17, etc.), hard and cartilaginous (7), etc. ; and it has been specially likened to morocco leather (6), to cracked glazed leather (11), to a sheet of parchment (5), and to the skin of boiled potatoes (13). It is cold to the touch.

The plates into which the integument is divided by the furrows and fissures differ greatly in size and shape. Some are quite large, and in certain instances the whole back has been covered by one enormous plate ; others are so small as to be little larger than pinhead-sized islands of epidermis ; and all the possible intermediate sizes may be found. The largest occur on the back, arms, hands, thighs, and feet, situations in which little or no movement occurs ; the smallest are met with on the head, anterior aspect of the thorax, and of the lower part of the abdomen, and round the anus and genitals. The plates are of almost every imaginable form ; they may be square, triangular, circular,

elongated, pentagonal, hexagonal, and so forth ; and the appearance produced has been compared by Radcliffe Crocker (53) to that of a "loosely-built stone wall." These epidermic islands have an average thickness of 4 or 5 mms. ; but they may measure as much as 7 or 8 mms., whilst on the other hand they may be as thin as tissue paper. The thickest are found on the back, scalp, and chest, and the thinnest on the hands and feet, and around the anus. The margins of the plates are generally bevelled off, and have a finely-fibrous appearance ; they terminate, however, occasionally in an abrupt manner. Sometimes there is a perceptible amount of imbrication. The surface of a plate is generally smooth to the touch, but on close inspection it is often possible to see that it is gently undulating, there being a series of alternating elevations and depressions. Along the line of the ridges are to be recognised whitish, punctiform, hollowed-out spots, which are found to be the dilated mouths of the sebaceous glands and hair-follicles, and here and there fine hairs are seen to project. If an epidermic plate be raised from the underlying cutis, it is seen that its attachment has been due to the interlocking of a series of conical projections (about 5 mm. in length), which exist both on the under surface of the squame and on the upper surface of the exposed cutis. The appearance thus produced has been compared to that of the rough tongue of the feline race. The attachment of the plates is less firm at the margins than near the centres, which are therefore somewhat depressed. In some cases it has happened that in certain areas of the body, as in the lower region of the abdomen and near the genitals, the plates have become detached, and a bleeding or moist red surface exposed. This detachment probably occurs during the extraction of the infant.

In exceptional cases small spines are developed on the plates ; thus, in No. 26 a row of these projections began on the forehead and extended over the head and down the back ; they were triangular in shape, from $\frac{1}{4}$ to $\frac{3}{4}$ in. in length, dark grey in colour, dense as horn, and resembled exactly the spines seen on a young alligator's back. This, however, would seem to be an unusual development in the cases which we are now considering.

The furrows, fissures, and rhagades have a somewhat sym-

metrical arrangement ; the two sides of the body are not markedly dissimilar. Their colour has been already described. They vary greatly in length, breadth, and depth. They may be little more than cracks ; they may, on the other hand, measure from 5 to 10 mms. in width. Some are quite superficial, and others pass right down to the cutis. The deeper ones are usually met with on the head in the neighbourhood of the ears. They pursue sometimes a straight, sometimes a wandering course, and are, of course, most numerous where the plates are present in the greatest amount. Many of them are covered by a thin, transparent pellicle which has been likened to a serous membrane, and which establishes a continuity of the epidermic surfaces (Thibierge) ; some, however, are quite destitute of any such covering, and from them a sanious, sanguinolent, or purulent fluid exudes which gives off a foetid or cadaveric odour. The former are no doubt old fissures which have skinned over and so become partially healed, whilst the latter are recent tears due to movements made during extra-uterine life.

It is now necessary to consider the special appearances met with in the various regions of the body, and the deformities of the different orifices produced by the thickened and inelastic state of the epidermis.

The head, which is often more rounded than usual, sometimes shows a fissure running backwards in the line of the sagittal suture. It is perhaps more frequent to find, as in No. 30, two fissures beginning near the anterior fontanelle and passing backwards, one on each side of the middle line, to end posteriorly by breaking up into a number of smaller cracks. In this way a large median line is produced on the vertex, and it is to its existence that much of the difficulty in diagnosis during labour is due. At the sides and back of the head the plates are more numerous. Some hairs are usually seen on the scalp, but they are short and small in number.

The region of the face is that in which the deforming effects of the cutaneous lesion are most evident (*vide* Plate III.). The plates at the sides of the face are usually larger than those near the middle line,—in fact, the latter are scales rather than plates.

A swelling in the parotid region has been noted in some cases (30, 40, etc.), giving to the cheeks a puffed-out appearance. The mouth is kept widely open by the contraction of surrounding parts, and from its angles radiate outwards fissures which have been aptly compared to the rhagades of syphilis (Thibierge). The buccal aperture is gaping, and the lips are thickened and everted so as to form a bright red or vermillion margin. This condition of eclabium is a very constant one, and hence it comes that the tongue and gums are unusually evident. The tongue is usually normal in appearance (it was described as bifid in No. 1); but both it and the gums, palate, and fauces were found intensely congested in No. 2. The chin is somewhat receding, and covered with small squames. In most cases a nose can hardly be said to exist (Schabel's specimen (16) formed an exception to this rule), and above the mouth and below the eyes there is usually nothing to be seen save two apertures surrounded by numerous small epidermic plates. The fact that the nose is not in relief is due to the absence of the nostrils and the external portion of the cartilaginous nasal septum. The two orifices which lead into the nasal fossæ are commonly blocked more or less completely by the hypertrophied epidermis.

Some of the older authors (*e.g.*, No. 1) thought that the eyeballs were absent, and that their place was taken by the two red fleshy masses that were seen to occupy the orbits; but it was soon shown that these fleshy tumours were really the greatly swollen and congested conjunctival surface of the everted eyelids. This condition of ectropion would seem to affect chiefly the upper eyelid, and it gives to the face a very hideous appearance. On separating the swollen lids the normal eyeball is revealed. The eyelashes are few in number or altogether absent, and the same remark applies to the eyebrows. The reddish mass formed by the everted lids becomes much more prominent when the infant cries. On the eyelids, as elsewhere, fissures and furrows are visible, and in some cases (*e.g.*, No. 5) wide and deep cracks pass outwards from the eyes into the surrounding integument.

The external ears seem to have disappeared entirely. Sometimes an opening lying amongst very thick and deeply fissured

epidermic plates can be made out leading into the auditory meatus. Sometimes there is no aperture, and the ear is represented by nothing more than a tubercle. In the less deformed cases the cutaneous fold marking off the auricle from the head has disappeared, and the cartilages of the ear are applied directly to the cranial bones.

The neck, which is unusually broad and thick, is often seen to be greatly denuded of its epidermic covering. This separation of plates occurs to a large extent during birth, and leaves a reddened moist surface exposed to view.

On the back the plates are large and the fissures few in number. In No. 16 the whole back, with the exception of the scapular regions, was covered by a single very large plate; in No. 30 there were two large islands of thickened epidermis in the position of the shoulders, while from the region of the neck a fissure passed downwards in the middle line to the upper dorsal area, where it divided into two branches passing forwards to join the lateral fissures on the abdomen, and formed the upper boundary of the largest plate on the body (11·5 cms. in vertical diameter, 30 cms. in transverse). In the case of Schab's first specimen (35) a small wandering furrow was found in the dorsal region pursuing a nearly median course, from which others passed outwards running almost parallel to the ribs to join the anterior mesial furrow; in the lumbar region there were scarcely any furrows, but parallel to the iliac crests were two grooves which united posteriorly at a point corresponding to the middle of the sacrum. The back in No. 17A had a somewhat peculiar appearance: there were a few fissures at the upper part, and the usual radiate arrangement of grooves at the lower; the intervening space showed none of these cracks, but in their place were some fifty-two pemphigoid bullæ containing a yellowish serum lying between the cutis and the epidermis, and varying in size from that of a pinhead to that of a threepenny piece.

On the anterior aspect of the thorax the fissures have in many cases (Nos. 5, 16, 31, etc.) a somewhat transverse arrangement, so that the plates of epidermis form bands; in other instances, however, there is an irregular furrow passing downwards nearly in the middle line; and in yet other specimens

(e.g., No. 7) the distribution of the cracks and grooves is quite irregular. Sometimes a vertical fissure passes down from the axilla to the iliac crest. There is no trace of nipples. On the anterior aspect of the abdomen the plates are usually smaller, and there are frequently areas from which the epidermis has separated. The insertion of the umbilical cord is commonly normal, and the thickening of the epidermis has not been traced on to the funis. The region immediately surrounding the navel is comparatively free from fissures.

In the case of female fœtuses the vulva is usually found to be in a gaping condition, exposing the hymen, and the labia majora are often enlarged and firmer in consistence than normal. In male infants the absence of the testicles from the scrotum has been occasionally noted ; sometimes the external genitals appear to have been arrested in development, and are represented simply by a tuberculated knob (7). Frequently the hardened epidermic layer is absent over the scrotum and its neighbourhood. The anus is sometimes normal in appearance ; occasionally it is narrow, and even imperforate ; it is not usually ectropic. There is usually a radiate arrangement of fissures round the anal orifice.

The condition of the limbs now calls for consideration. They usually retain the intra-uterine attitude of flexion, and have often a swollen appearance as if oedematous ; and whilst they are sometimes traversed by longitudinal fissures, a circular arrangement of clefts is more commonly met with. The fissures are most numerous on the flexor aspect of the limbs, in the axillæ, and on the groins, and the cracks are specially deep round the wrists and ankles. The hands and feet are greatly thickened and deformed ; they are sometimes clubbed (*talipes varus*). The digits are curiously deformed, and sometimes resemble birds' claws (*onychogryphosis*), sometimes are very small in size (even absent), and sometimes are united to each other.

3 Internal Appearances.—A complete dissection of the ichthyotic fœtus does not seem to have been often made ; but the best descriptions of the internal appearances are found in connexion with Nos. 17, 25, 30, and 40. If it be permissible to

draw conclusions from such a small number of autopsies, it may be stated that as a general rule the viscera show nothing abnormal save an unusual degree of congestion. The mucous membranes are not at all thickened. An external cephal-hematoma was found near the anterior fontanelle in No. 25, on the right parietal bone in No. 40. The bones of the cranium are normal in size and form, but the sutures and fontanelles cannot be palpated on account of the thickened epidermis. There was no change in the brain or its membranes in No. 25; in No. 30 the cerebrum was congested, the cerebellum and medulla were healthy, and there were sub-arachnoidal haemorrhages; and in No. 40 the brain was congested, whilst the meninges were normal. In No. 17 there was localized pleurisy on both sides, and the corresponding portions of lung tissue were hyperæmic and atelectatic, whilst the other parts were somewhat emphysematous; in No. 25 the right lung was oedematous and the left was in parts atelectatic; in No. 30 the lungs, especially immediately below the pleura, were here and there in an unexpanded state; and in No. 40 there was slight subacute pulmonary oedema. In No. 33 the cause of death was widespread broncho-pneumonia. In No. 17 the heart was described as normal, but both the foramen ovale and ductus arteriosus were open, although the infant had lived for nine days; in No. 25 the pulmonary semilunar valves were slightly fenestrated, but the heart and great vessels were otherwise healthy; in No. 30, in which the child lived seven days, the only cardiac anomaly was the patent condition of the foramen and ductus; and in No. 40 there was nothing unusual in the state of the heart, but the thymus was voluminous.

The abdomen contained from 60 to 100 grammes of reddish fluid with some clots in No. 30, but there were no traces of peritonitis. Congestion of the liver was noted in No. 17; of the liver, spleen, pancreas, and kidneys in No. 25; of these organs, and also of the adrenals, in No. 30; and of the spleen, kidneys, and liver in No. 40 (the pancreas and supra-renal capsules being in this instance normal). The mucous membrane of the intestinal tract had a healthy appearance in No. 25; but in No. 40 there was some enlargement of the follicles and Peyer's patches in the

lower part of the ileum. The intestine contained meconium or yellow faeces according to the shorter or longer term of life. In No. 25 the internal genitals (female) were normal; but in No. 30 the uterus was larger than usual, and the left broad ligament was abnormal. In No. 40 the left testicle was in the scrotum; the right lay midway in the inguinal canal.

II. *Microscopic Characters.*

Not many of the reported cases of foetal ichthyosis are accompanied by good descriptions of the microscopical appearances of the integument and internal organs; but Nos. 12, 16, 17, 24, 25, 30, 32, 33, 34, 35, and 40 form exceptions, for in them the histology of the cutaneous structures at least is studied in detail. Before proceeding to consider the typical lesion as manifested by the microscopical changes in the integument, I may briefly enumerate the observations that have been made upon the minute anatomy of the viscera.

Kyber (25) found no increase in the interstitial tissue in the liver, kidneys, or spleen, and no thickening of the mucous membrane of the stomach or intestines; there was, however, considerable congestion of all the internal organs. In Livingstone's specimen (30) the liver, besides being congested, showed fatty degeneration of its cells; the kidneys and spleen, save for the congestion, were normal in their microscopical appearances; and as regards the lungs, the examination of the expanded portions revealed a normal epithelium in the bronchi and air-vessels, whilst in many of the alveoli were crowds of red blood corpuscles and large numbers of round bacteria. Finally, Carbone (40) examined the liver, spleen, kidneys, the mucous membrane of the mouth, pharynx, and larynx, and the eyeballs, and found in all of them normal microscopical appearances.

We may now pass to the study of the minute anatomy of the integument, and upon this the best investigations are those of Kyber and Carbone. The following description is based mainly upon the conclusions arrived at by these two observers (25 and 40) (*vide* Plate II.).

Carbone found that the appearances were different in different parts of the body-surface, and he described, first, the conditions

DESCRIPTION OF PLATE IV.

Microscopic Appearances of Skin of Palm of Hand in Kyber's Case
of Fœtal Ichthyosis.

a, Stratum corneum with sweat canals; *b*, stratum Malpighii; *c*, projection passing down between the papillæ; *d*, sweat ducts; *e*, sweat glands.

PLATE IV.



found in a typical epidermic plate ; second, those in the cicatricial areas or furrows ; and, third, those in the recent fissures or rhagades. Even with the naked eye it can be seen that the thickened horny layer shows numerous orifices destined for the passage of hairs ; these passages are particularly numerous on the scalp, and give to it the spongy appearance noted by Müller. Round the orifices the epidermic cells are arranged concentrically, resembling in some degree the cell-nests of epitheliomata.

(1.) If a section be made of any of the horny plates covering the surface of an ichthyotic foetus, it will be found that its most striking feature is enormous thickening of the epidermic layer. In Carbone's specimen the epidermis varied in thickness from 2 mms. on the scalp and 1 mm. on the neck to about '3 mm. or '5 mm. on the rest of the body. This increase in the epidermis is due entirely to the hypertrophy of the stratum corneum, for the rete Malpighii, with the exception of what will be immediately described as the interpapillary prolongations, is not only not thickened, but is even in some places diminished in thickness.

To begin with the cells of the rete Malpighii, those which lie nearest to the cutis, and which have in the skin of a normal foetus a regular columnar shape, have not in specimens of foetal ichthyosis always so regular a form. They tend to be lower, as if compressed, and in the places where there is a special increase in the stratum corneum they may even be cutical or rounded in shape. Lying upon this basal layer of cells are two or three strata of polygonal cells, having a normal appearance and arrangement, and showing between them the normal intercellular spaces.

There is no stratum granulosum of Langerhans to be recognised : there is no layer of flattened cells with nuclei surrounded by granules of eleidin (Ranvier) or kerato-hyalin (Waldeyer), the substance which occupies an intermediate position between protoplasm and keratin. This layer is also absent in normal foetuses of the age to which most specimens of ichthyosis belong. Carbone, however, in his case noticed some kerato-hyalin bearing cells in the external root-sheath of the hairs.

There is, on the other hand, a well-marked stratum lucidum of Schrön, a layer in which the cells are more or less flattened

and have in greater or less degree lost their distinct outlines, and in which the nuclei are few in number, have their long axis parallel to the surface of the epidermis, and are surrounded by large perinuclear cavities. These nuclei are shrivelled, and their chromatin contents have condensed into granules or fused into a small body. The stratum lucidum, which is a homogeneous stratum about to become keratinised, consists in a normal fœtus of little more than a single layer of cells ; but in foetal ichthyosis, whilst it varies considerably in different places, it is always composed of several rows of flattened cell elements.

The stratum corneum is, as has been already stated, enormously thickened in foetal ichthyosis. Further, in this disease the passage from the stratum lucidum into the horny layer is not sudden and sharp, as in normal fœtuses, but is more gradual. There are first several layers of horny cells, greatly flattened, and containing nuclei which are scarcely visible, for they do not stain more deeply than the surrounding tissue ; then come several layers of still more flattened and condensed cells, forming an almost homogeneous stratum, with a very fine striation, resembling in appearance the epidermis of the adult palm. This gradual transition is not always to be seen ; the stratum lucidum sometimes passes directly into a horny layer as compact and dense as nail. The stratum corneum is not, however, in all places so compact as has been described : where to the naked eye the epidermis has a fibrillated appearance, it is seen by the microscope that the horny layer is irregularly divided into lamellæ running in various directions and separated from each other by large empty spaces. Another difference which exists between the stratum corneum in the normal and in the ichthyotic fœtus consists in its reaction with osmic acid : in the former this reagent stains deeply the superficial and the deepest layers of cells, leaving the intermediate layers unstained, whilst in the latter there is no such colour reaction, or only the presence of some fine black lines. Carbone attributes this to impeded sebaceous secretion.

It is now necessary, before considering the state of the skin-glands and hairs, to return to the study of the rete Malpighii. In the nuclei of the deeply situated cells of this layer Carbone looked carefully for signs of karyokinesis, but was able to find

DESCRIPTION OF PLATE IVA.

KYBER'S SPECIMEN OF FœTAL ICHTHYOSIS.

FIG. 1.—Vertical Section of the Skin of the Chest in a thickened area.
a, Stratum corneum, with hair canals containing lanugo hairs ;
b, stratum Malpighii ; *c*, sweat glands ; *d*, hair sac ; *e* and *f*, seba-
ceous glands filled with fat cells ; *g*, lanugo hairs ; *h*, corium.

FIG. 2.—Vertical Section of Hair Sac with Sebaceous Gland from Skin
of Head (Kyber).

FIG. 3.—Vertical Section of Skin of Palm of Hand in a Normal Infant
(Kyber). *a*, Stratum corneum ; *b*, stratum Malpighii ; *c*, inter-
papillary projections ; *d*, corium ; *e*, sudoriparous glands ; *f*, adipose
tissue.

FIG. 4.—Transverse Section of Hair Sac containing Hair from Skin of
Head (Kyber).

Fig. 1.

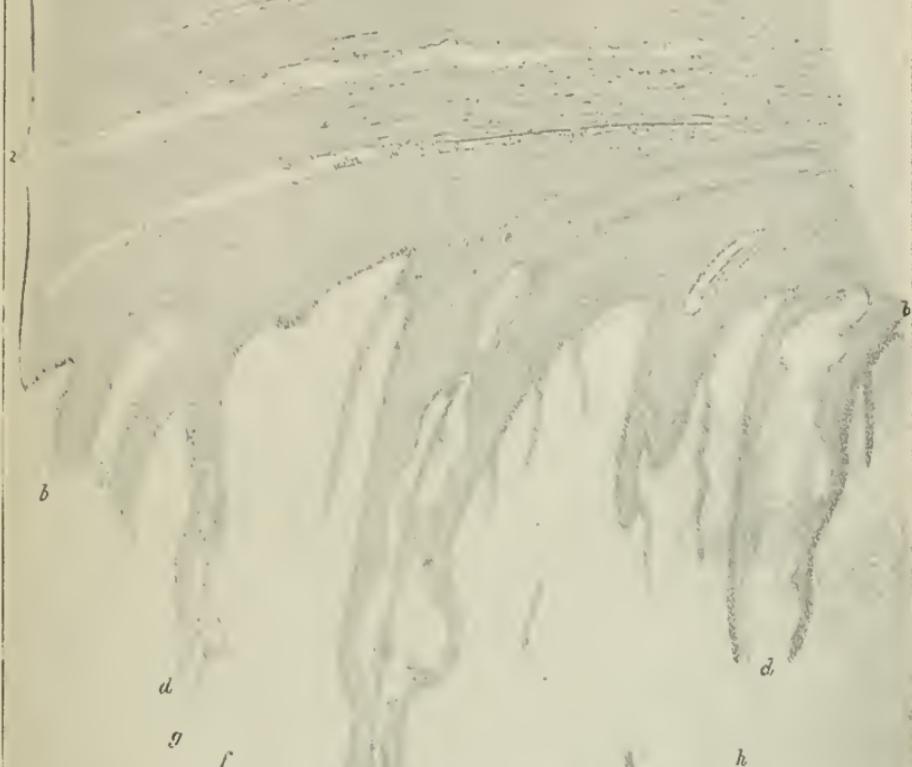


Fig. 2.

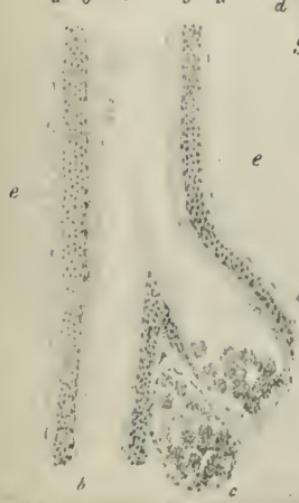


Fig. 4.



Fig. 3.

very few mitotic appearances. He therefore concluded that the proliferating activity of these cells was not only not increased, but was actually diminished, and he was on this account confirmed in the belief that the Malpighian layer was really less in thickness than normal. Now Kyber (25) had regarded it as increased in thickness and possessed of greater proliferating activity. Carbone, however, refers to Kyber's own drawings, which show that the rete Malpighii, save in the interpapillary processes, is diminished rather than increased in thickness. These interpapillary processes have a deceptive appearance: they seem to be enlarged, for they pass more deeply into the underlying cutis between its papillæ; but whilst they are elongated vertically they are diminished in thickness, and are therefore, according to Carbone, really less in size than normal. The interpapillary processes are specially long and thin in the skin of the palm of the hand; and in that region, as well as on the sole of the foot, there is another curious appearance to be seen. The normal lines due to the folding of the digits are absent, and in their place are found large downward prolongations of the horny layer, having a conical form with base uppermost, and being sheathed by a thin Malpighian layer, which sends out interpapillary processes into the surrounding tissue. These cone-like prolongations of the stratum corneum are also traversed by many sudoriparous gland ducts having a direction parallel to the long diameter of the cones. Kyber explains the formation of these horny wedges in the following way: when the hand is flexed at the metacarpo-phalangeal joints it is found that the skin surfaces bounding the groove thus formed are brought into contact; if this, then, happen at an early period of fœtal life, and if the flexion be long-continued, the contiguous skin surfaces will fuse together, and there will be formed just such a horny wedge in the derma as has been described; further, since the ducts of the sudoriparous glands pass through these cones in a direction parallel to their axis and perpendicular to the surface of the skin, it is probable that the folding of the skin occurred at a time anterior to the formation of the sweat glands, for if it had occurred later the ducts would have had a direction perpendicular to the sides of the cones.

It is now necessary to consider the condition of the *hair follicles*, the *sebaceous* and the *sudoriparous glands*. The stratum corneum sends down a prolongation into the mouth or funnel-shaped portion of the hair follicles, enormously dilating it. In some cases the mouth of the follicle may be completely closed by the horny substance, in others it contains a mass of broken-down epithelium and sebum. The follicles that contain lanugo hair show this plugging most markedly. The external root-sheath of the hair is also greatly thickened (it is almost five times thicker than normal, according to Carbone) and in part keratinised ; and whilst in the case of the hairs of the head this thickened sheath rarely passes further downwards than the point at which the sebaceous gland opens into the follicle, in the case of the lanugo hairs it extends to a considerably lower level than that. The internal root-sheath may be described as normal. The hairs themselves are, according to Kyber, abnormally thin. The sebaceous glands attached to the hairs of the head are usually atrophied ; they are lined with low cubical cells and contain normal polygonal fat-bearing cells, and rarely does the horny layer of the external root-sheath penetrate into their ducts. Elsewhere, however, there is usually this penetration of the sebaceous gland duct, and in some places the process of keratinisation may go so far as to convert the whole gland into a cavity surrounded by completely horny walls.

Kyber (25) states that the sudoriparous glands are hypertrophied, and that their ducts are longer both in the portion that traverses the corium and in that which passes through the stratum corneum. Carbone, however, failed to find this hypertrophy, although he noted that the glands were well developed.

The *corium* or *cutis vera* does not, in foetal ichthyosis, show any marked departure from the normal. It has been stated by some (*e.g.*, 16) that the papillæ are hypertrophied ; but whilst it is quite true that they show a projection upwards corresponding to the projection downwards of the horny interpapillary processes, they are also narrower, and are probably not really increased in size. There is in some places an increase in the number of connective tissue elements and leucocytes ; the blood-vessels have been regarded as not markedly abnormal.

With regard to the *subcutaneous tissue*, no important changes have been noted ; but the amount of adipose tissue contained in it is usually less than normal, and this diminution in the fat is specially marked in the palm of the hand and sole of the foot (Kyber).

(2.) The histological appearances revealed by an examination of the integument in the cicatricial areas or furrows differ somewhat from those which have been described as characteristic of the plates. The epidermis is not so much thickened : the stratum corneum is notably thinner than in the case of the plates, but is still thicker than normal, and the rete Malpighii may be made up of only two or three rows of flattened cells. If a complete section be made of one of these furrows, it will be seen how the horny layer, which is thick at the sides where it becomes continuous with that on the plates, dips down, and is reduced greatly in thickness on the floor of the furrow. Sometimes, according to Schab (35), a little projection is seen at the middle of the furrow, which is perhaps the remains of an interpapillary epidermic cone. Both the papillæ of the cutis and the interpapillary processes of the rete Malpighii are little developed in the cicatricial areas, and in their central parts the hairs and sebaceous glands are not to be found, although some sudoriparous glands may be present. In the parts adjacent to the furrows hairs are found which pass obliquely upwards and outwards, diverging from the margin of the grooves. The direction of these hairs, as well as the absence of such appendages in the middle of the furrow, is probably due to the fact that the laceration of the skin, of which the furrow is the cicatrix, took place before the development of the germs of the hair follicles. The cutis vera has a condensed appearance.

(3.) The microscopical appearances presented by a recent fissure or crack have been thus described by Carbone (40). Usually the crack extends down to the cutis vera, upon which lie some remnants of the rete Malpighii, covered with crusts formed of pus cells, red blood-corpuscles, and broken-down epithelial cells. The papillæ seem to be infiltrated with leuco-

cytes, and in them are seen greatly distended bloodvessels and sometimes small haemorrhages.

In some places the fissure is not so deep ; the stratum lucidum is torn through, but the rete Malpighii, infiltrated with leucocytes, is left in position. Sometimes traces of the horny layer of the external root-sheath of the hairs may be found. The deeply-lying portions of the hair follicles are always preserved, and this accounts for the occasional presence of hairs in the cicatrical areas or furrows.

Such are the microscopical appearances of foetal ichthyosis as described by Kyber and Carbone, and, stated shortly, they consist in enormous thickening of the stratum corneum with the projection downwards of the interpapillary processes, in an extension of the keratinising process to the sebaceous gland and in part to the external root-sheath of the hairs (the internal root-sheath remaining normal), and in possibly some degree of hypertrophy of the sudoriparous glands and of the rete Malpighii. The cutis vera is practically normal, whilst there is a diminution in the amount of fat in the subcutaneous tissue. The slight discrepancies observed in the descriptions given by Kyber and Carbone are probably due to the fact that the case examined by the former was a more marked one than that seen by the latter. With regard to the thickness and proliferative activity of the Malpighian layer, Carbone himself admits that it seems necessary to concede that at some stage in the morbid process this stratum had these characteristics, although in his specimen the stage had been passed. This conclusion is rendered more probable by the fact that Jahn (17) in his case found the cutaneous papillæ notably hypertrophied ; and not only were they hyperæmic, but they showed in some places a state of evident inflammation. Schabel (16) also noted hypertrophy of the papillæ.

With regard to the chemistry of the epidermic scales, Livingstone (30) found that they were composed of fat, cholesterine, and of what might be hippuric acid ; whilst their burnt residue was made up of salts of lime, magnesia, and iron.

B. MORBID ANATOMY OF THE FœTAL ANNEXA.

Unfortunately little or nothing is known of the state of the fœtal annexa in cases of fœtal ichthyosis. The placenta and umbilical cord were described as normal in Nos. 17, 25, 26, and 30; but the cord was noted to be very thin in Nos. 7, 14, 15, and 17A; and in No. 7 the placenta was said to have been small, soft, and easily torn.* The thickening of the epidermis does not seem to have extended to the covering of the cord. It was noted in Nos. 17 and 26 that the liquor amnii existed in large amount, and in the former instance it was said to be normal in appearance. This is somewhat surprising; it would seem more natural for the liquor amnii to be deficient rather than excessive in amount. A thorough investigation of the placenta, cord, membranes, and liquor amnii in cases of fœtal ichthyosis is much to be desired.

* The placenta and membranes were described as normal in Sherwell's case (41A).

CHAPTER VII.

FŒTAL ICHTHYOSIS (GRAVE TYPE)—Continued.

ETIOLOGY; PATHOGENESIS; DIAGNOSIS; PROGNOSIS; TREATMENT; FŒTAL ICHTHYOSIS IN THE LOWER ANIMALS; LITERATURE.

ETIOLOGY.

ALMOST nothing can be affirmed with regard to the etiology of foetal ichthyosis, for practically nothing is known of the causes which give rise to this intra-uterine malady. Like many other foetal diseases, it has been ascribed to maternal impressions, to syphilis in the parents, and to heredity ; but it would seem that the only thing that is certain about its etiology is that the condition is not due to any one of these causes. In the recorded cases the absence of syphilis is generally conclusively proved ; and it is also almost universally stated that the parents and other relatives were entirely free not only from similar cutaneous affections, but also from all skin eruptions. In fact, the father and mother seem always to have been both healthy and strong ; and it cannot be considered that Jahn's case (17), in which there was maternal rickets (or osteomalacia), and in which, therefore, we find an exception to this rule, forms a serious argument against the exclusion of heredity as an etiological factor. Sex seems to be of no importance whatever, for half the cases were females and half males.

There are, however, two facts which have a possible bearing upon the causation of the morbid state which must not be overlooked. The first is the more important, and it is the circumstance that in some cases the same mother has given birth to more than one ichthyotic foetus. Of the 39 mothers who gave birth to infants with this disease, three at least (*i.e.*, 7·6 per cent.) bore more than one similarly affected. In Okel's case (14, 15) the mother first had a normal infant and then two

ichthyotic foetuses in two successive years ; and in Houel's case (24) there were also two infants suffering from this malady born to the same parents. Oestreicher's case (37, 38, 39) is specially interesting, for in this instance a woman had three healthy infants by her husband, and then after his death gave birth to three ichthyotic foetuses in succession by another man. Foetal ichthyosis is a very rare disease ; and it is, to say the least of it, very striking that this "*family prevalence*" should have been present so often. We do not yet know much about the significance of family prevalence, but it is a phenomenon that has been noticed in foetal maladies other than ichthyosis. It may be that hereditary tendencies in the parents which could not when acting singly produce the malady, are able to do so when united in the fertilized ovum. Oestreicher's case especially seems to favour this view.

The second fact is somewhat allied to the above, and has to do with the blood-relationship of the parents. It is true that the existence of consanguinity was only noted in one case ; but in that instance (40), it was of a very close kind, for the parents were uncle and niece. These two facts must not be forgotten in relation to the possible hereditary origin of foetal ichthyosis.

PATHOGENESIS.

It is now necessary to consider the nature and possible mode of origin of the condition known as foetal ichthyosis, and the probable time in intra-uterine life at which the morbid process begins.

I. Two chief theories of the *nature of this pathological condition* have been advanced. According to the one, it is ichthyosis occurring *in utero* and manifesting itself in a more intense form than is seen in extra-uterine life ; according to the other, it is not a kind of ichthyosis, but a general seborrhœa sicca or squamosa. Before proceeding to consider these two views, I may state briefly that some of the older writers regarded it as a monstrosity or deformity (1, 3, 5, and 46), and that one (Hinze, 2) looked upon it as a variety of lepra Græcorum ; but it is unnecessary to discuss these views, for the latter is untenable, and

as to the former, it would be difficult indeed always to fix a hard and fast line between diseases and deformities in foetal pathology.

Sir James Y. Simpson (43) seems to have been the first to emphasize the view that the disease was of the nature of ichthyosis, although he may in part have been led to this conclusion by the fact that Grætzer (42) discussed the condition along with ichthyosis hystrix. Further, Simpson appears at one time to have thought that it was related in some degree to the "skin-bound" disease (sclerema); but in 1843 he said that he had abandoned this idea, and had arrived at the conclusion that it appeared to be "much more analogous to ichthyosis than to be any other skin disease that can be referred to." He explained the differences which existed between it and the ordinary forms of ichthyosis by the difference in the environmental conditions under which it was developed. Simpson's theory of the nature of the disease and the name which he gave to it were practically accepted by all those who wrote prior to 1876. In that year F. Hebra (45) gave the great weight of his authority as a dermatologist to the view that so-called foetal ichthyosis was really general seborrhœa, and stated certain objections to the ichthyosis-theory. The writers who followed Hebra were greatly influenced by his opinions; and generally abandoned Simpson's theory, either using the terms ichthyosis congenita or ichthyosis sebacea, with an apology for their inaccuracy, or adopting the expressions keratosis (Lebert) or keratoma, names which, while expressing the nature of the morbid change, did not bind them to either the ichthyosis- or the seborrhœa-theory. Of late years, however, there has been in many quarters a reaction against the dictum of F. Hebra and in favour of the view originally stated by Simpson. This reaction has been greatly aided by the arguments of Thibierge (49); by the authority of Auspitz, Caspary, G. T. Elliot, Hans Hebra, Hallopeau, Crocker, and others; and by the occurrence of cases which must be regarded as the connecting links between the grave form of ichthyosis foetalis and ichthyosis vulgaris of adult life.

It may, therefore, be well at this point to consider the chief arguments that have been brought forward against the ichthyosis-theory and the answers that can be given to them.

The first objection taken by Hebra (45) to the ichthyosis-theory was founded upon the absence of any evidence to show that the morbid process in the foetus was ever transformed into the ichthyosis vulgaris of childhood and adult life. Now, since death ensued in every case during the first ten days of life, it is, of course, impossible to say what changes the disease might or might not have undergone had the child lived. It must, therefore, be admitted that up to the present time no case has been recorded in which an infant born with the grave form of foetal ichthyosis has lived long enough to demonstrate the conversion of this type into the ordinary variety of adult ichthyosis ; but cases of a milder kind* have been met with, have lived, and have shown various changes, some in the direction of complete or more commonly partial cure, and others in the direction of the establishment of ordinary adult ichthyosis. (It is in this latter category that I should place the case noted by Hebra, in which he says that an infant born with so-called foetal ichthyosis was with care kept in life, and showed many years afterwards a perfectly healthy skin.) The occurrence of these milder forms of foetal ichthyosis and the changes which have been traced in them considerably weaken the force of Hebra's first objection.

In the second place, it was stated that ichthyosis, although a *congenital* malady in the loose sense of the term, was never developed before the second year of life. It is now known, however, that this opinion is erroneous ; for, leaving out of account at present the "attenuated form" of foetal ichthyosis which may be present at birth, and which will be described in the next chapter, cases of ichthyosis have been noted which have developed within the first year, and, indeed, during the early months of life, although they have not become very conspicuous till the second year (Crocker). It may be, as H. Hebra (48) suggests, that the rare occurrence of ichthyosis in infancy is due to the prolonged soaking which the skin has had from the liquor amnii and to the daily bathing to which it is subjected during the early months of extra-uterine life ; it will appear when the effect of the former has worn off, and when the latter has been in great measure discontinued.

* *Vide Chapter VIII.*

The third objection that has been brought against the ichthyosis-theory is the absence of any proof of the heredity of the foetal condition. Now, in the first place, the hereditary nature of adult ichthyosis cannot always be proved : as Radcliffe Crocker (53) says "it is in many cases, but by no means in all, hereditary." Second, although there is no proof that foetal ichthyosis may be sometimes transmitted by heredity, this does not necessarily mean that it cannot be so transmitted. For it must be remembered that no ichthyotic foetus has long survived its birth, and there has, therefore, been as yet no opportunity for a hereditary tendency to manifest itself. But it is possible to go further than this, for an indication of the occasional hereditary nature of foetal ichthyosis does exist in the occurrence of "family prevalence" in some cases. No great weight, therefore, can be attached to the third objection.

In the fourth place, it has been stated that so-called foetal ichthyosis and ichthyosis vulgaris cannot be the same disease, because their clinical features and morbid anatomy differ so widely. Now, it must of course be granted that there is a marked difference between a typical instance of foetal and one of adult ichthyosis ; but this difference is in great part explained if it be assumed that the former shows a much more advanced stage and a much graver type than the latter. *In utero* there is not that constant friction and movement which we may suppose are instrumental in limiting both the degree and the extent of the disease in the adult, and therefore it is quite reasonable to expect a universal distribution and an extensive cutaneous lesion in foetal ichthyosis. Further, the morbid process is essentially the same in both cases ; it is a hyperkeratosis in the adult as well as in the foetal malady. The difference is really one of degree, not of kind. All the peculiar features of foetal ichthyosis can be accounted for on this hypothesis: the thickness of the scales is due to the greater accumulation of epidermic cells ; the furrows and fissures are to be explained by the absence of such elasticity in the skin as will permit its expansion to make room for the rapidly growing skeleton ; and the deformities of the limbs, mouth, eyes, nose, and ears are caused by the arrested development of these parts in consequence of the constricting effect of

the horny cutaneous covering. In foetal ichthyosis, it is true, the rete Malpighii and papillæ are not often in a hypertrophic condition; but the same thing has been occasionally noticed in adult ichthyosis by Kaposi (52) and Boegehold (*Virchow's Archiv*, lxxix. p. 545).*

Whilst these arguments are sufficiently strong to greatly diminish the force of F. Hebra's fourth objection to the ichthyosis-theory, still stronger are forthcoming. In recent years several cases of what has been called the "attenuated form" of foetal ichthyosis have been observed, and in them one can hardly fail to see the connecting link between the typically severe variety which has been discussed and ordinary adult ichthyosis. Nothing further need here be said concerning these cases, for they will be fully considered in the next chapter. It is, however, necessary to refer now to the two very interesting cases (sister and brother) reported by Lang (54), for they demonstrate in an unexpected way the possibility of the development of what has always been regarded as true foetal ichthyosis *after birth*. In the first, the infant, a female, was born at the 39th week of pregnancy, and was healthy, but rather small and stunted. At the third week of life it was noted that the skin was stiff and shiny, and nodes formed, which burst and allowed pus to escape. At the fourth week ectropion of the eyelids (especially the upper) was observed, and at the age of one year the child became blind. The left eye was "phthisical," the right showed a corneal ulcer. Lang saw the patient when she was two years and a quarter old. The skin was as shiny as satin. There were ragged epidermic lamellæ (the size of a pea or a bean, or even larger, and as thick as tissue paper) with raised borders and firmly attached and depressed centres, which could be more or less easily separated as squames or scales. The external ears had a smaller vertical diameter than normal, and posteriorly they were distorted. In the neighbourhood of the anus the skin was radially arranged in thick folds. There was no panniculus adiposus. The skin of the limbs was contracted so that the fingers, which were badly developed and too short, were flexed at all the joints. The nails

* An interesting paper on the histology of ichthyosis is that by S. Giovannini, *Arch. f. Derm. und Syph.*, xxvii. p. 3, 1894.

were twisted. There was retraction of the skin in the popliteal space. Immediately behind the toes, on the heel, and at the radio-carpal joint, the parts seemed to be constricted by the epidermic plates. The scales were distributed all over the body (face, limbs, and trunk), but the contraction of the skin was confined to the limbs, for the integument of the trunk was described as sufficient, and felt like a thin sheet of parchment with stiff folds. Bathing twice daily and rubbing the parts with olive oil loosened the plates, but the child died in four weeks, and at the autopsy, ichthyosis, rachitis, atrophia bulbi sinistra et nervi optici sinistri, hypertrophy cordis, hydrothorax, hydropericardium, hydrops ascites, and bronchitis were found. In the second child, a male and the brother of the above patient, the first changes in the skin were noticed at the second month, and were of the same kind as in the little girl. The same treatment was adopted, and three weeks afterwards it was noted that the child could sleep with closed eyes, which had been impossible before on account of the ectropic condition of the eyelids. After about five weeks of such treatment the patient was removed from the hospital, and died at home some two months later at the age of a little over one year. Now, Lang's cases are especially important, for they demonstrate that a condition precisely similar to foetal ichthyosis, and having the same deforming effect upon eyes, ears, and limbs, may be developed in extra-uterine life.

From all that has been said it is, I think, permissible to draw the conclusions that the use of the name foetal ichthyosis is perfectly justifiable, and that the malady is essentially the same in nature (although different in degree) as adult ichthyosis. All the signs of a hyperkeratosis are present, and none of a hypersteatosis. At the same time there can be no possible objection to the employment of the phrase "keratosis diffusa epidermica," if that be preferred.

II. The possible mode of origin of the morbid process in the skin may now be briefly considered. It was suggested by Löcherer (22) that an excessive formation of sebum and epidermis took place, which hindered the exfoliation of the stratum

corneum, and set up an irritation which produced a hypertrophy of the papillæ of the derma. Carbone (40), however, objects that in his case the process probably began before the formation of the sebaceous glands. It seems necessary to suppose that there is at some time or other excessive activity of the cells of the rete Malpighii ; but of the way in which this activity is set up nothing can be definitely said. Barkow (17A), however, makes an interesting suggestion. He believes that there are two distinct stages in the development of foetal ichthyosis. The first of these, which he calls the pemphigus-stage, and which is characterised by the formation of bullæ, has usually passed away before the birth of the infant, but may occasionally be observed, if carefully looked for, in localised areas of the body surface. The epidermis covering the most distended bullæ bursts and allows the serous contents to mix with the liquor amnii. The blebs collapse ; but the tears remain, or ultimately unite to form a much thinner epidermis. Between the fissures are the plates of undivided skin with thickened epidermis and hypertrophied papillæ. This hypertrophy constitutes the second stage of the morbid process, and is left behind when the primary inflammatory condition has passed away or has given rise to serous exudation and bullous formation.

It has, again, been suggested by some that ichthyosis originates in a tropho-neurosis. Thus Leloir * ("Contribution à l'étude des affections cutanées d'origine trophique," *Arch. de Physiol. norm. et path.*, p. 391, 1881) found lesions of atrophic degenerative neuritis in the cutaneous nerves and spinal nerve roots (especially the posterior) in some cases of general ichthyosis dating from birth, and he quotes Eulenburg and Arnozan as giving each a case due to chronic neuritis ; but with regard to foetal ichthyosis all that can be said is that Carbone (40) carefully looked for such lesions in his specimen and found them not. We are forced to conclude, with the last-named writer, that the process is a homologous hyperplasia of the epidermis whose mode of origin is not less unknown than that of general hyperplasia of adipose or of osseous tissue.

* *Comptes rendus hebdom. des Séances de l'Acad. des Sciences*, lxxxix. p. 1123, 1879; and xci. p. 134, 1880.

III. It is possible to give a more satisfactory answer to the question, When does the process usually begin *in utero*? It is generally believed (Nos. 25, 30, and 40) that it cannot begin at an earlier date than the third month of intra-uterine life, whilst its commencement is not usually delayed beyond the beginning of the fourth month. Kyber (25) was of opinion that the normal development of the central nervous system and of the crystalline lens of the eye proved that the change in the epiblast did not originate in the early weeks of foetal life; whilst he thought that the arrangement of the hairs in the fissures, and of the sweat-ducts in the skin of the palms and soles, along with the amount of extension of the healed fissures on the head, showed that it had already begun in the fourth month. For, according to Kölliker, the development of the hairs takes place at the end of the third or the beginning of the fourth month *in utero*. Carbone (40) also, from the absence of hairs in the centre of the cicatrices, and from the arrangement of the sweat glands and interpapillary processes of the rete Malpighii, comes to the conclusion that foetal ichthyosis begins at some time between the third and fourth months of intra-uterine life.

IV. With regard to the mode of origin of the characteristic features of foetal ichthyosis, it is also possible to advance a plausible theory. If it be granted that at an early period of foetal life the normal distensible epidermis is replaced by a horny inelastic covering, then it can be conceived that as the skeleton and internal viscera go on rapidly growing, this hard and unyielding investment must rupture at certain places in order to allow of expansion. This tearing will take place where the skin is most apt to be put upon the stretch by movements, etc.,—as, for example, on the anterior aspect of the trunk, the flexor surface of the limbs, especially at the joints, and on the scalp. In this way are produced the fissures, whilst the furrows are simply partially healed fissures. With regard to the ectropic condition of the eyes and mouth, these deformities must be ascribed to the tension of the surrounding skin, which is so great as to prevent the closure of these orifices. The stunted condition of the hands and feet, and the want of development of the nose

and ears, are to be explained by the restraining influence upon growth exercised by the horny investing coat of altered epidermis. In these parts of the body fissures do not seem to occur so often or so readily.

DIAGNOSIS.

It is unnecessary to say anything with regard to the diagnosis of fœtal ichthyosis of the grave type *after birth*. The external appearance of the infant is so striking, and so unlike that in any other malady, that there can be no hesitation in coming to a right conclusion as to its nature. It may, however, be readily imagined that *during labour* the condition of the skin of the presenting part may lead to misconception and error. The breech or other part may easily be mistaken for the head, and the head itself may be thought to possess some anomaly of its osseous framework, such as anencephaly (7).

PROGNOSIS.

Cases of fœtal ichthyosis of the grave type have invariably proved fatal to the infant,* and death has ensued at a time varying from a few hours to nine days post-partum. Barkow's case (17A), it is true, is said to have been dead-born, and F. Hebra (45) states that he saw a patient recover (unfortunately he gives no details); but these are the only exceptions to the rule that an ichthyotic fœtus is born alive but survives only for a few hours or days. The cause of death is no doubt complex: the infant is originally weak, for it is usually born prematurely, and this weakness is rapidly increased by insomnia (due to pain on movement), by inability to suck, by hindrance to respiration, and by suppuration in the various cracks and fissures, which soon become "the haunts of pyogenic microbes;" further, there is the interference with the functions of the skin due to the horny investing layer, although recent investigations have tended to show that this is a less potent cause of death than has been supposed; and, finally, there are the visceral congestions,

* Sherwell (41A) has recently reported a case in which an infant affected with fœtal ichthyosis was still alive at the age of five months; but the descriptive notes show that the disease was of moderate severity.

and especially inflammatory states of the lungs and pleura. From one or other, or from all these causes, death ensues ; but it may yet be possible that at some future time, and with improved or novel therapeutic resources, infants affected with this grave type of ichthyosis may be saved.

TREATMENT.

As will be readily understood, no method of treatment has proved efficacious in this disease. The two chief therapeutic indications must be to soften and remove the hard epidermic covering and to maintain the strength of the infant. For the first purpose warm baths have been employed, along with the use of ointments, of vaseline, lanoline, glycerine, and the like. Antiseptic washes (*e.g.*, boracic lotion) may be used to prevent suppuration occurring in the fissures, and oil or ointments may be rubbed into the lips and surrounding skin to soften them, and so enable the infant to suck. In order to carry out the second indication the child must be fed with a spoon or per rectum, and all the necessary details as to clothing, warming, etc., strictly carried out.

It is a matter for regret that no writer has described the state of the thyroid gland in ichthyotic fœtuses ; but even in the absence of such information it might be well to try feeding with thyroid extract in any cases which may in the future be met with.

FŒTAL ICHTHYOSIS IN THE LOWER ANIMALS.

It is an interesting fact in comparative pathology that a condition exactly similar to the foetal ichthyosis of the human subject is occasionally met with in fœtuses of some of the lower animals. Cases of this kind in the new-born calf have been met with by Gurlt (55), Charcot (56), Liebreich (57), Luschka (58), Harpeck (59), Goubaux (61), Bland Sutton (62), and others.

It is not necessary to detail the pathological characters of these specimens, for they were in all points similar to those met with in the human fœtus. With regard, however, to the history of the cases, one of two facts may be noted. Thus, it is recorded in connexion with Harpeck's observation that a healthy cow

gave birth first to an ichthyotic calf, then at the next parturition had a calf whose skin, indeed, was normal, but which was monstrous (absence of head and extremities), and finally bore another ichthyotic animal. The occurrence in the lower animals of the phenomenon in heredity known as family prevalence (which has been already observed in the human subject) is noteworthy. The phenomenon, however, was exceptional, for in Gurlt's first case (55) the cow had on a previous occasion had a healthy calf. Liebreich's observation was interesting in another direction : in the estate of Sachsendorf thirty-six cows had for a long time been impregnated by the same two healthy bulls and had always borne normal calves, but between the years 1850 and 1852 four ichthyotic animals were put into the world, each of which came from a healthy mother that had on no previous occasion had an abnormal calf-fœtus ; during these two years, however, it had been noted that the younger bull had showed a diminished sexual instinct, and Liebreich was of opinion that the fault did not at any rate lie with the mother animal. This suggestion of a possible paternal etiological factor in foetal ichthyosis of the calf has some interest when considered in conjunction with Oestreicher's cases (37, 38, and 39) in the human subject. Foetal ichthyosis in the calf, as in the human fœtus, seems always to have proved fatal soon after birth.

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CHAPTER VIII.

FŒTAL ICHTHYOSIS (MILD TYPE).

DEFINITION; SYNONYMS; HISTORICAL NOTES; FREQUENCY; CLINICAL HISTORY AND SYMPTOMATOLOGY; PATHOLOGY; PATHOGENESIS; DIAGNOSIS; PROGNOSIS; TREATMENT; LITERATURE.

BESIDES the grave form of foetal ichthyosis which has been described in the two preceding chapters, there is a variety of the disease which must, I think, be regarded as the mild or attenuated form; and to it attention must now be directed.

Definition.—The mild type of foetal ichthyosis may be defined as a skin disease of the foetus, characterised at birth by the presence of a continuous layer of a collodion-like substance over the whole body, and later by the desquamation of this substance in the form of small flakes resembling pieces of tissue paper, accompanied sometimes by ectropion and eclabium, and not usually proving fatal to life, but occasionally terminating in complete or partial cure.

Synonyms.—Seeligmann (1) found it difficult to group this disease with any known cutaneous malady, and thought it was unnecessary to give to it any name other than "*epidermic desquamation in the new-born infant.*" Gould (2 and 3) and Fröbelius (4) called their cases specimens of "*congenital ichthyosis,*" and their example was followed by Gidon (7), Weisse (12), Fox (13), and Schab (24). Auspitz (5) preferred the term "*ichthyosis neonatorum,*" whilst Caspary (16 and 17), and Munnich (21 and 22) used the expression "*ichthyosis fœtalis.*" Perez (8) named his case one of "*general sclerosis of the skin in the new-born,*" and Behrend (15) entitled his communication "*Ein Fall Idiopathischer Angeborener Haut-atrophie.*" The other writers, however, used the word "*ichthyosis*" in one or other combina-

tion: "*intra-uterine ichthyosis*" (Elliot, 25 and 26), and "*ittiosi sebacea dei neonati*" (Tommasoli, 20). Hutchinson (6), however, called his specimen one of the "*pityriasis variety of congenital Xeroderma*," whilst Alexander (14) entitled his "*a case for diagnosis*." Another synonym, perhaps the best, is that used by Hallopeau and Watelet (28), "*forme atténuee de l'ichthyose fœtale*." A popular expression is "*Alligator-boy*" (13), and a semi-popular one might be "*collodion fœtus*."^{*} I have used Hallopeau's expression in the slightly altered form of *mild type of fœtal ichthyosis*.

Historical Notes.—The first observation which can be identified as one of the mild form of fœtal ichthyosis was that reported in 1841 by Seeligmann (1) in his inaugural dissertation. Gould's two cases (2 and 3), in which umbilical haemorrhage and jaundice were also present, must, I think, be looked upon as examples of the same disease; they were published in 1854 and 1855. So were also the cases of Fröbelius (4) and Auspitz (5) reported in 1865 and 1869 respectively. The patient whose history was given by Hutchinson (6) in 1875 may with some degree of probability be regarded as one of congenital ichthyosis. Gidon (7) noted a case in 1879, and Perez (8) one in 1880. Of the three examples seen by Warner (9, 10, and 11) in 1882, the third was undoubtedly a typical example of the attenuated form of fœtal ichthyosis. Weisse's case (12) led to an interesting discussion in which several American dermatologists took part; and Fox's patient was the "*Alligator-boy*" of the "*Dime Museum*." Alexander (14) showed a male infant to the New York Dermatological Society in 1884, which led to an interesting discussion: Fox thought it was an instance of congenital ichthyosis, Piffard regarded it as *dermatitis exfoliativa neonatorum*, and Bronson also looked upon it as a parakeratosis similar to Ritter's disease. Behrend (15), in 1885, described a typical case under the name of *idiopathic congenital atrophy of the skin*; but to Caspary (16 and 17), who in the same year described two brothers affected with this malady, much credit is due for the

* Fournier, in the course of a discussion upon Hallopeau's specimen, suggested the name "*desquamation collodionnée*."

convincing way in which he proved that the disease is really ichthyosis. Other cases were soon afterwards reported by Michelson (18 and 19), Tommasoli (20), Munnich (21 and 22), Schwimmer (23), and Schab (24), between the years 1886 and 1890. Elliot, in 1891 (25 and 26), described two patients (sisters) with intra-uterine ichthyosis of the mild type, and gave a useful summary of the previous observations. Hallopeau and Watelet (28), by their report of a case to the French Dermatological Society, excited an interesting discussion in which Thibierge, Besnier, Vidal, and Fournier took part.

Frequency.—If any opinion may be formed from the number of reported cases, it would seem that the mild type of foetal ichthyosis occurs with about the same degree of frequency as the grave form. From the year 1841 to the present time thirty-seven cases of the latter kind have been recorded and thirty-three of the former.* Possibly, however, as the existence of the mild or attenuated form becomes better known it will be found that it is of commoner occurrence than the grave type.

CLINICAL HISTORY AND SYMPTOMATOLOGY.

It will be convenient to consider in order, first, the maternal ; second, the paternal ; and third, the family history ; and thereafter to take up the symptoms of ichthyosis as met with in the patient himself. Since this mild form of the disease is much less fatal to life than the grave type, it will be possible to give a much fuller account of its symptomatology.

A. *Maternal History.*—In a considerable number of the recorded cases little or no information is forthcoming with regard to the health of the mother ; but in a few instances there is a somewhat complete account. In No. 5 the mother was 30 years old when she gave birth to an ichthyotic infant, and she was 28 in No. 24. It is not always stated how many children she had had ; but she was a i.-para in No. 13 ; a ii.-para in Nos. 14 and 24 ; and a iii.-para in No. 1. In No. 5 the mother

* The bibliographical list at the end of this chapter would seem to show only twenty-nine, but in certain of the records it was incidentally stated that another infant had been similarly affected.

had been six times pregnant; her third and fourth children were ichthyotic, and her sixth gestation ended in the birth of twins, one of which had the disease. In No. 11 six healthy infants were born before the one with ichthyosis and three after it; and in No. 12 two older children were healthy and one younger, the ichthyotic child being the third. In Behrend's case the tenth infant and an earlier one both had the disease. Nos. 16 and 17 were the only children of the same mother, and both were ichthyotic; so were Nos. 25 and 26. The mother of Michelson's patients (18 and 19) had one other child. The occurrence of more than one ichthyotic infant in the same family was therefore frequently observed.

The mother's health was usually good. It was specially described as such in Nos. 1, 11, 13, 14, 16 and 17, 21 and 22, and 25 and 26; but in No. 5 the mother had frequently suffered from pruritus, especially during her pregnancies. In all the other cases it may be taken for granted, in the absence of any information to the contrary, that the mother was free at least from any skin disease.

The pregnancy which resulted in the birth of an ichthyotic infant sometimes terminated prematurely: at the 7th month in No. 15; at the 8th month in Nos. 11, 13, and 22; at the 9th month in Nos. 1 and 24; and "prematurely" in Nos. 16 and 17. In Nos. 7, 25, and 26, however, the gestation went to the full time. In the other cases no reference is made to this fact. In some instances (1, 13, and 24), an "impression" during pregnancy was alleged by the mother as the cause of the infantile disease. These "impressions" were as follow: the sight at the 7th month of a child in the desquamative stage of scarlet fever in No. 1; the sight of an alligator and of a dog in a fit in No. 13 (at 4½ months); and a fright and the sight of a fish in No. 24. In most of the recorded cases the pregnancy was uneventful; but in No. 5 the mother suffered from vomiting, various congestions, and pruritus; and in No. 24 the foetal movements were violent. The pregnancy was plural in No. 5, but the twins were not of the same sex; the ichthyotic one was a male, the normal one a female.

Details of the labour were seldom forthcoming: it was easy

and normal in Nos. 3, 14, 16, and 17, delayed in No. 1, and tedious and irregular in No. 13. With regard to the puerperium, it was stated in No. 5 that the pruritus from which the mother had suffered in pregnancy disappeared three weeks after her confinement.

B. *Paternal History*.—In many cases no mention is made of the condition of the father; but in Nos. 1, 5, 11, 12, 13, 14, 16, 21, 24, and 25, he was a healthy man. In Nos. 11 and 16 the parents were blood-relations (cousins). In No. 25 the father had been twice married; by his first wife he had several healthy children, and one (a girl) with hyperkeratosis limited to the palmar surface of the fingers; by his second wife he had two children, both ichthyotic.

C. *Family History*.—In none of the recorded cases of the mild type of foetal ichthyosis is there any note of the occurrence of this disease, or of ichthyosis vulgaris in the ancestors of the patient. The only indication, therefore, of heredity in connexion with this malady is to be found in the occurrence of "family prevalence" in the cases seen by Gould (2 and 3), Auspitz (5), Hutchinson (6), Behrend (15), Caspary (16 and 17), Michelson (18 and 19), Munnich (20 and 21), and Elliot (25 and 26). In No. 11 the mother and her sister married their cousins (two brothers), and each woman gave birth to an ichthyotic infant. It is to be noted, however, that in No. 5 (plural pregnancy) only one of the twins was affected.

D. *Infantile History and Symptomatology*.—Since in most of the cases of the mild type of foetal ichthyosis life was prolonged for some months or years, it is possible to give a fuller description of its symptomatology than of that of the grave variety. It will be convenient to describe, first, the clinical features seen at birth, and, second, the changes which occur in later life.

I. *Clinical Features at Birth and during first week of life.*

(a.) *Sex of the Infant*.—In Nos. 4, 8, 15, 20, 27, and 28, I have been unable to find a record of the sex of the infant; but in fourteen cases (Nos. 3, 5, 6, 9, 10, 11, 12, 13, 14, 16, 17, 21, 23, and 29) it was a male, and in nine cases (Nos. 1, 2, 7, 18, 19, 22, 24, 25, and 26) a female. In No. 5 two other ichthyotic infants

are incidentally referred to,—one a male, the other a female ; and in No. 6 there is the notice of another male. If these be added to the others, it is seen that there have been sixteen males and ten females (in the proportion of 100 to 62·5).

(b.) *Size and Weight.*—Since in several of the cases the infants were born prematurely, it is not surprising that they were occasionally described as puny or small (Nos. 2, 5, 12, 13, 15, and 24). The weight was 3 lbs. in No. 15 ; 4½ lbs. in No. 13 ; 5½ lbs. in No. 2 ; and 2550 grammes in No. 24. In No. 3, however, the child weighed 9 lbs., and in some other cases it was described as of the usual size. In the great majority of the records these details were not given. It is probable, however, that a full-time ichthyotic infant is not much below the average in size and weight.

(c.) *Vitality.*—In no case was the infant born dead ; but in a few instances it died in a week or two ; thus in No. 24 it lived only three days, in No. 3 twelve days, and in No. 2 sixteen days. Life lasted for three months in No. 29. In No. 3, however, the death was due to causes other than the ichthyosis, and in No. 7, in which the infant lived for eleven months, there was broncho-pneumonia. It cannot be said, therefore, that the mild form of foetal ichthyosis is fatal, either to intra-uterine or to extra-uterine life. When last seen by the observer, the age was seven days in No. 28 ; one month in No. 14 ; six weeks in No. 8 ; three months in No. 5 ; nine months in No. 22 ; seventeen months in No. 15 ; eighteen months in No. 16 ; three years in Nos. 1 and 10 ; four years in Nos. 9, 17, and 21 ; five years in No. 13 ; seven years in No. 26 ; eight years in No. 25 ; nine years in No. 11 ; ten years in No. 12 ; thirteen years in No. 19 ; twenty years in No. 23 ; twenty-one years in No. 18 ; and sixty-one years in No. 6 (a doubtful case).

(d.) *State of the Integumentary System.*—In all the recorded cases the skin presented an abnormal appearance at birth, but in some instances (Nos. 1, 3, 13, and 25), it was only after the thick layer of "vernix caseosa" had been removed by the first bath that the condition characteristic of this variety of ichthyosis became apparent. The whole body is covered with a firm, dry, shining, and tense membrane. This membrane has been de-

scribed as parchment-like (Nos. 1, 5, and 15); but it has been more customary to state that the skin looked as if it had been covered with a layer of collodion (Nos. 7 and 28), or of amber coloured (14) or brownish varnish (25), or of "poudre de riz" (28). Sometimes it formed a whitish pellicle (28), at other times a dusky brown, or reddish brown, or brownish yellow skin (Nos. 5, 15, and 24). Warner stated that in one of his cases (No. 11), the integument looked as if it had been scalded. Hallopeau and Watelet said that the surface of the body of their patient (28) seemed as if it had been immersed in a bath of tincture of iodine, and Auspitz (5) compared the skin to that of a duck's feet. The membrane is tightly stretched upon the surface of the body ["fest wie ein Trommel," says Behrend (15)], and its dry, glistening, or waxy appearance has often been emphasized. Perez (8) spoke of the infant as covered with a horny cuirass, an "ongle immense."

In a short time (varying from a quarter of an hour (28) to several days) cracks and fissures begin to appear in this collodion-like layer, and soon thereafter desquamation or exfoliation commences. In some instances these fissures may have been present at birth, but even then their number is small at first, and is rapidly and greatly increased when the infant begins to make active movements. Most commonly they are quite superficial, but sometimes they are deep bleeding rhagades (*e.g.*, No. 2). The cracks vary in number, and usually follow the lines of the natural skin folds. They are often specially numerous round the joints and on the face and head, and the first to appear are generally those near the mouth. In this way the epidermis covering the whole body is divided into irregular areas, varying in size, and in these areas desquamation of the cuticle takes place. Sometimes there is a separation of large yellow or brown squames and lamellæ; at other times small fragments like films of white tissue, silk, or satin paper exfoliate. The dry or slightly moist surface left exposed by the desquamation is usually of a lighter colour than the detached epidermis, but it soon becomes darker, and cracks rapidly begin to appear in it. In this manner a continuous process of exfoliation goes on all over the body. In No. 1 fragments of the size of

the palm of the hand, and carrying numerous hairs, sometimes fell off in the region of the scalp, and left exposed a new epidermis with small hairs. In some cases (Nos. 4 and 13) the surface areas are covered, not with a thin pellicle, but with a thick horny plate, and in appearance approach, therefore, very closely to typical ichthyosis in the adult ; but this is exceptional. Sometimes the infant seems to have suffered from considerable pruritus, as in No. 1, but of course this symptom may have been frequently present and have been overlooked or unrecognised on account of the age of the infant. Hairs seem usually to have been absent, but occasionally they were seen on the scalp.

It may be well here to mention certain deformities which are undoubtedly due to the state of the integument. These are of the same kind as in the grave type of fœtal ichthyosis, but are less severe. The mouth is often held half open by the contracted state of the surrounding skin, and suckling is therefore difficult, if not impossible, until the cracks and fissures occur and allow some freedom of movement. The nostrils are commonly blocked by the epidermic membrane, which may, indeed, have to be removed before respiration can be satisfactorily established (No. 1). The external ears are often glued, as it were, to the scalp by this same membranous covering, and the auditory meatus may be blocked. Occasionally the eyelids are in a state of ectropion, but this may not, in other instances, be developed till the infant is older. The limbs are usually more or less fixed in the foetal attitude of flexion, the tense condition of the skin not permitting freedom of movement at the joints. The digits have an imperfectly developed appearance, and are covered by skin like that of a cadaver. The fingers are sometimes (12 and 16) drawn to the ulnar side of the hand ; they are also flexed and claw-like, and the nails may be gryphotic (onychogryphosis). The toes also have been described (15) as short, warty stumps with claw-like nails.

(e.) *Symptoms in the other Systems.*—The chief symptoms which are associated with this form of fœtal ichthyosis are : more or less inability to move the limbs ; difficulty in suckling (child could not be put to the breast till eight days old in

No. 1); respiration difficult, or superficial and frequent (No. 24); and immobility of the facial muscles, sometimes preventing crying. The bowels are usually regular, the urine normal, and micturition easy. In No. 3, in which there was icterus from imperforate bile ducts, the stools were like putty and the urine was jaundiced. Sleep is sometimes good; at other times it is interrupted by pruritus. The appetite is usually healthy; but swallowing is in some cases attended with difficulty. Hearing and sight may be defective.

II. *Clinical Features developed after Birth.*

With regard to the condition of the skin, this, in some cases, becomes normal in a "few" weeks, as in No. 4, or in six weeks, as in No. 8, whilst in other instances there is only some improvement in the severity of the lesion, as in Nos. 5 and 14; but usually the morbid state continues unchanged up to the time of observation, and in one case (No. 6) desquamation of the cuticle was still present at the advanced age of 61 years. In Nos. 25 and 26 involution of the disease had occurred in certain parts of the body at the age of eight and seven years respectively, and in these cases, curiously enough, the involution took place in the very parts of the body (face, neck, and limbs) which are usually the parts of predilection in the adult form of ichthyosis.

The after-history of these cases does not properly fall within the province of this work, so I shall only give an account of the progress of the malady in one or two typical cases.

In Seeligman's patient (No. 1) the skin, which at birth was covered by a dry, shining, tense membrane, soon began to show larger and smaller cracks near the joints and elsewhere. Desquamation then began in long and broad or in short and narrow pieces, and was never afterwards absent. This desquamation was greater, the mother thought, at the time of the new moon. It occurred over the surface of the whole body, but was most marked on the head, where a piece the size of the palm of the hand, with all the hairs attached, would sometimes separate. There was great pruritus. At the age of six months, on various parts of the body were seen white maculae with a red areola and a yellow punctate centre. The epidermis at these places was

somewhat raised, the yellow spot increased, and a small tumour formed, from which (on being opened) yellow fœtid matter exuded. These puriform discharges occurred at intervals on various parts of the body. The lymphatic glands of the neck, axillæ, and groins swelled. *Tinea capitis* appeared, the abdomen became greatly swollen and hard, and along with these scrofulous signs photophobia was developed. The appetite for food was good, the bowels were regular, the urine was normal and was expelled in large amount. The skin was always dry, and sleep was usually good, save when interrupted by pruritus.

The girl at three years was of a size equal to her age. The head was somewhat large in comparison to the rest of the body. The slightly yellow hairs of the head were agglutinated by crusts. Some traces of eyebrows could be seen. The upper and lower eyelids of both eyes were in a state of ectropion, and most of the eyelashes of the upper eyelid were absent. The upper lip looked upwards; but the rest of the face was normal. The conchæ of the ears were normal in size, but were very rigid and hard. From the left ear a fœtid discharge always issued. The abdomen was distended and hard to the touch. The limbs could not easily be moved; the fingers were somewhat curved, and the toes were very small and imperfect. The skin all over the body had the appearances usually met with in typical adult ichthyosis.

Caspary's patient (16), a boy of 18 months, did not look older than 6 months. There was marked ectropion; and eclabium of both lips, but especially of the upper, existed. The skin everywhere was atrophic and exfoliating. The middle third of the cornea was exposed during sleep, and showed a leucomatous change. The very small nose was not exactly deformed, but the nares were plugged with dried-up secretion. The auricles were drawn backwards tightly against the head; the upper half was almost absent, the lower was thickened. From the crack-like auditory meatus on the left side pus issued. The skin appeared to be universally retracted to the maximum, and was covered by large and small scales. Here and there were small abscesses in the cutis, and behind the left ear was a large collection of pus in the subcutaneous tissue. The cutaneous scales were typically ichthyotic; they were few in number on the back of head, posterior

aspect of the trunk, and on the nates. There were signs of pruritus (scratch-marks) on the arms and elsewhere. The joints were in a flexed position. The fingers were bent to the ulnar side, but neither they nor the toes were strikingly altered. The nails were convexly curved and were not shining. There were traces of eyelashes and eyebrows ; under the scales on the scalp were a few lanugo hairs, but none elsewhere. The anus was almost at the level of surrounding parts. The fontanelles were closed, and there were no signs of rickets. The muscular development was poor, and there was scarcely any panniculus adiposus. There was no perspiration at any time. All the bodily functions, except sight and hearing, were normally performed.

In one of G. T. Elliot's cases (25) there were the usual appearances at and soon after birth. At the age of three years there was diminution in the intensity of the process on the face and limbs ; the horny lamellæ were still formed, but became thinner ; and in eighteen months the skin on these parts became soft, smooth, and natural. At the age of eight years the girl was well grown and showed no malformations. She was bright and intelligent, but slightly anaemic. She never perspired, save on the upper lip and limbs. The bodily functions were well performed. On the affected parts there was typical ichthyosis. The hair on the head was abundant, glossy, and natural ; there was a good development of lanugo on the arms and legs. She had measles,* which ran a mild course ; but with it there was a return of the ichthyosis to the face and limbs, disappearing again on treatment with salicylic acid. Measles, therefore, would seem to have caused a temporary reversion to the original ichthyotic state. In some cases the cold was felt very much.

From the record of these three cases some idea will have been formed of the course which foetal ichthyosis takes in after life.

* In Gidon's case (7) varicella occurred at the eighth month. The appearance of the eruption evidently gave great pain to the child. Each pustule elevated the epidermic layer and was surrounded by a bright red ring. Recovery from the varicella ensued ; but the infant died from broncho-pneumonia at the age of eleven months.

PATHOLOGY.

Macroscopic Appearances.

The morbid condition of the skin in this form of fœtal ichthyosis has been already described (*vide Symptomatology*) in so far as it is visible to the naked eye during the life of the patient. Little here remains to be said upon the subject, for most of the patients were living when seen by the authors who described them ; and in Nos. 2, 3, and 24, in which the infant died soon after birth, nothing of note is added to the account already given of the macroscopic appearances of the skin. With regard to the condition of the internal organs as seen by the naked eye we have little information ; in No. 24 it is simply stated that no direct cause of death was found at the autopsy ; in No. 2 the brain was hydrocephalic ; and in No. 3, in which umbilical haemorrhage and jaundice were present during life, the liver was very dark in colour, engorged with blood, and friable in consistency ; the gall-bladder, which was flaccid, contained about a teaspoonful of clear fluid like synovia ; and the umbilical vessels were pervious, while the cystic and common bile-ducts were not. A complete account of an autopsy upon an infant dying of this disease is still, however, a desideratum.

Microscopic Appearances.

What is known of the microscopic anatomy of the skin has been learned from the examination of small portions of the diseased integument removed during the life of the patient. In Nos. 7 and 14 it is simply stated that the exfoliated scales consisted of epidermic cells and amorphous matter. In No. 24 a piece of skin removed from the forearm showed that the stratum corneum was not much thicker than normal ; the rete Malpighii was of proportionate extent in the interpapillary areas, but above the papillæ of the cutis vera it was markedly thinned.

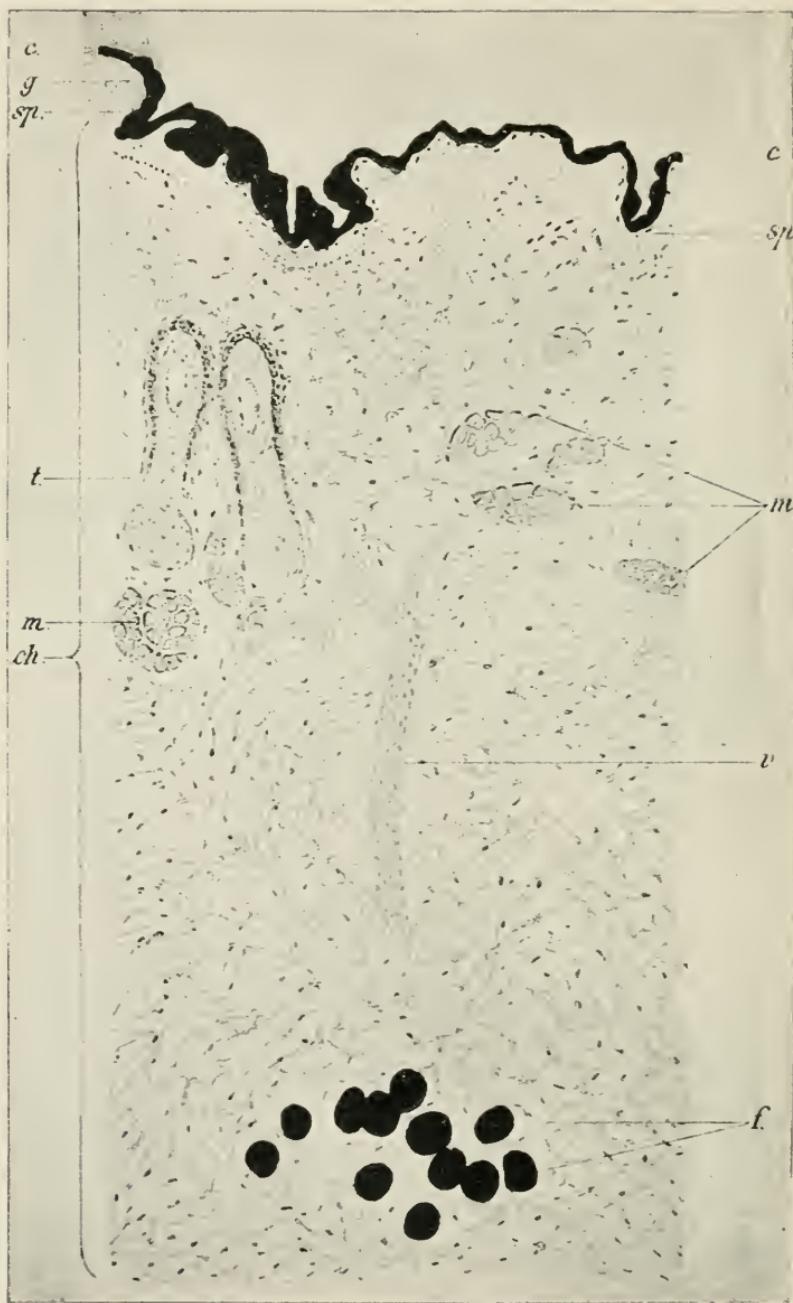
The best description of the histology of the skin was that given by Caspary (No. 17). He cut a piece of skin from the abdominal wall near the umbilicus, and having treated it with Flemming's solution, made microscopic sections of it and compared them with those of the skin of a normal (somewhat

atrophic) infant. He found that the ichthyotic skin had only half the thickness of the normal, and noted also that the panicularis adiposus was much reduced. The epidermis, however, was relatively increased in the case of the ichthyotic infant, for it constituted fully one-quarter of the total skin thickness. There was no black-stained surface layer such as is found on normal skin (treated with Flemming's solution); this was doubtless due to the absence of fat. Both the stratum corneum and the stratum lucidum as well as the rete Malpighii were increased in thickness, and even the stratum granulosum was somewhat thicker than normal (the child was 18 months old at the time). Both the cylindrical and the prickle cells of the rete had their usual characters. The cutis vera consisted of a wide-meshed network of poorly developed connective tissue, with scarcely any elastic fibres. The papillæ were not really increased in size, for their greater height was counterbalanced by their diminished breadth; the increase in their vertical measurement was due to the dipping down of the interpapillary rete Malpighii. The cutis vera was poorly supplied with blood. No sebaceous glands were seen in any of the sections, and there were only a few hair follicles with lanugo hairs. There was no indication of the smooth musculature of the corium or of the arrectores pilorum. The sudoriparous glands, however, appeared to be unusually well developed. The appearances found by Caspary indicated hypertrophy of all the component parts of the epidermis, along with atrophy of the cutis vera and of the subcutaneous adipose tissue. The well-developed state of the sudoriparous glands, however, was noteworthy. It cannot, of course, be affirmed that all cases of this form of foetal ichthyosis will have the same histological appearances. Probably differences will be found in the state of the cells of the rete Malpighii, etc., just as in adult ichthyosis vulgaris and hystrix. In the absence of other observations, however, Caspary's description must, for the present, be regarded as typical (*vide* Plates V. and VI.).

ETIOLOGY.

Little can be safely affirmed with regard to the etiology of this form of foetal ichthyosis. Certain facts may, however, be noted.

PLATE V.



DESCRIPTION OF PLATES V. AND VI.

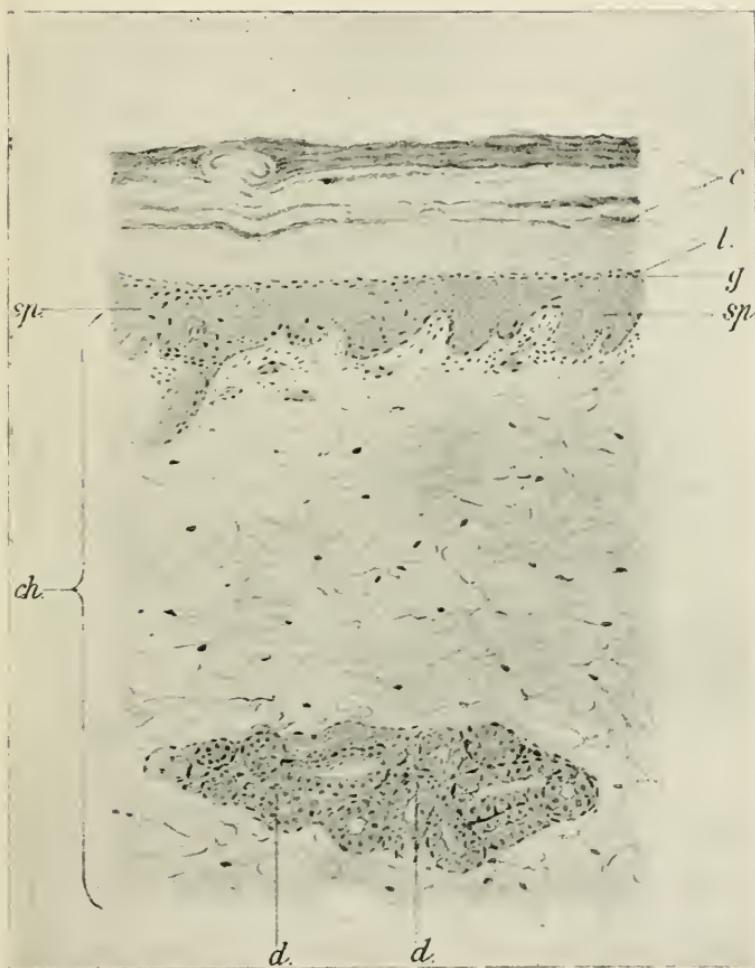
PLATE V.—Vertical Section of Normal Skin of Abdomen in an Atrophic Infant, $1\frac{1}{2}$ years old (Caspary).

PLATE VI.—Vertical Section of Skin of Abdomen in an Ichthyotic Infant, $1\frac{1}{2}$ years old (Caspary).

In both Plates the letters have the same signification.

c, Stratum corneum; *l*, stratum lucidum; *g*, stratum granulosum; *sp*, stratum spinosum; *ch*, corium; *d*, sudoriparous glands; *f*, fat cells; *m*, transversely cut bundle of non-striped muscle fibres; *t*, sebaceous glands; *v*, vein.

PLATE VI.



(1.) The first is the freedom of the parents and their immediate progenitors from this form, and, indeed, from any form of skin disease, for the fact that in one case (5) the mother suffered from pruritus during the pregnancies which ended in the birth of ichthyotic infants cannot by itself have much weight.

(2.) The second noteworthy circumstance is the frequent occurrence of more than one ichthyotic infant in the same family. Of the twenty-four mothers who gave birth to children with this disease, eight (or one-third) had more than one so affected (Nos. 2, 5, 6, 15, 16, 18, 20, and 25). In another instance (No. 11) the mother had only one ichthyotic foetus, but her sister had also one. Family prevalence may therefore be taken as proved for this disease; but it must at the same time be remembered that in one case (5) a woman gave birth to twins (probably binovular) of which only one suffered from ichthyosis.

(3.) That consanguinity in the parents has any etiological importance is doubtful; it was, at any rate, noted only in two cases (11 and 16).

(4.) A possible paternal influence is suggested by the history in No. 25; but it is a solitary instance.

(5.) The etiological value of the "maternal impressions" recorded in Nos. 1, 13, and 24 is extremely problematical.

(6.) The sex of the infant cannot be regarded as of much causal importance; in sixteen of the recorded cases the child was a male, and in ten a female.

All that can safely be affirmed is that heredity is the most powerful etiological factor; but the heredity is of the peculiar kind known as "family prevalence," the exact value of which has not yet been determined.

PATHOGENESIS.

The nature of this disease can hardly be doubted. It is ichthyosis occurring in the foetus and persisting after birth; but all the writers who have described cases have not been of this opinion.

Seligmann (1), for instance, pointed out that the disease resembled both psoriasis and ichthyosis, but thought that it was

neither the one nor the other. He considered that it was not psoriasis, because that skin affection never covered the whole body, and did not give rise to pruritus. It was not ichthyosis, for there were signs of inflammation present, and the epidermis was not condensed to form imbricated scales. He thought it best to regard it simply as an epidermic desquamation of the new-born, due probably to a chronic inflammation of the skin. Hutchinson (6) was inclined to look upon it as the congenital form of xeroderma ; but in reality there is no hard and fast line between mild ichthyosis and marked xeroderma.

In the discussion that followed the reading of Weisse's case (12), Taylor stated his belief that it was not ichthyosis but pityriasis rubra. In another discussion, however—that upon Alexander's communication (14)—Taylor adopted for it the name of congenital keratosis, whilst Piffard took up the opinion that it was dermatitis exfoliativa neonatorum. Behrend (15) regarded it neither as an inflammatory condition nor as a new formation, but as an idiopathic skin-atrophy. He, however, thought that it might be similar to Hutchinson's "pityriasis variety of congenital xeroderma," and showed how it could be distinguished from scleroderma, xeroderma pigmentosum (Kaposi), and eczema squamosum. On the occasion when Hallopeau and Watelet (28) showed their patient to the Société française de Dermatologie et de Syphiligraphie, Vidal stated that he had seen similar appearances in some cases of foetal syphilis; but Fournier said that he had never seen anything like it in syphilis, for in that malady there was a maceration or moist desquamation, not "*une desquamation collodionnée*," as in Hallopeau's case.

Now, whilst it cannot be denied that the attenuated form of foetal ichthyosis resembles in some ways dermatitis exfoliativa and congenital syphilitic affections of the skin, it has seemed, in the opinion of most authorities, to have closer affinities to ichthyosis than to any other cutaneous malady. All that was said in favour of the ichthyosis-theory when speaking of the grave type may be repeated here, and, indeed, some of the arguments apply with greater force to the attenuated than to the more marked form. With regard, for instance, to the

question of heredity, "family prevalence" has been more often demonstrated in the former than in the latter disease. Then, again, the pathological appearances in the attenuated form show less dissimilarity when compared with those in ichthyosis vulgaris than do those in the grave foetal type. In fact, the mild form may be said to occupy, in this respect, an intermediate position between grave foetal ichthyosis and ichthyosis vulgaris in the adult: it differs from the former in showing less advanced changes in the epidermis, and from the latter in the presence of the associated deformities of the eyes, nose, mouth, ears, and limbs. Further, it has been clearly shown that the attenuated form may develop into ordinary ichthyosis in childhood and adult life, in some instances persisting as a marked xeroderma, in others becoming changed into ichthyosis vulgaris or cornea. The microscopic appearances do not prevent the acceptance of the ichthyosis-theory, and the atrophic changes that have been emphasized by some writers are doubtless secondary phenomena. Finally, Lang's cases, which have been already referred to (*vide* Chap. VII.) serve the important purpose of showing that all the features of foetal ichthyosis may be developed after birth.

With regard to the mode of origin of this form of foetal ichthyosis, nothing can be said with certainty; indeed, the pathogenesis of even the adult form is still most obscure. To say that it originates in a congenital anomaly in the development of the skin, and especially of its stratum corneum, is simply to put what we know already into other words, and does not further our knowledge of its evolution. The same may be said of the assertion that it is a hyperkeratosis or a parakeratosis. It seems probable that at some time or other in intra-uterine life there must be an inflammatory affection of the skin, possibly of the rete Malpighii; but this statement can only be made as an unproved hypothesis. It is possible that it begins at a later date in foetal life than does the grave type.

DIAGNOSIS.

The diagnosis of the attenuated form of foetal ichthyosis can scarcely be difficult: the presence of a collodion-like layer covering the whole body of the infant at birth, the subsequent

appearance of fissures and cracks, the splitting up of the thickened epidermis into scales and flakes, and the associated deformities, make up a clinical picture so characteristic that it can hardly be mistaken for anything else.

From *xeroderma* it is not necessary rigidly to distinguish it, for that disease in a most marked form runs insensibly into the milder types of ichthyosis.

From *xeroderma pigmentosum*, *atrophoderma pigmentosum*, or *dermatosis Kaposi* it is differentiated by the fact that it (ichthyosis) is present at birth and does not show pigment spots, telangiectases, etc.

The absence of desquamation in *scleroderma*, along with the fact that it has never yet been seen in the new-born infant, should serve to diagnose this malady from foetal ichthyosis.

Psoriasis is not universal in its distribution, and has not been seen before the age of eight months. It begins also in the form of papules, which become capped with white scales.

Eczema squamosum is rare in early childhood, and the history of the case, the distribution of the lesion, etc., will serve to distinguish it from congenital ichthyosis.

Fœtuses affected with *syphilis* sometimes show certain resemblances to those suffering from ichthyosis; but the desquamation in the former is of the moist rather than of the dry type, and the history of the case, along with other things, will usually serve to differentiate the two diseases. The absence of any syphilitic history is a most marked character of the clinical record in all cases of foetal ichthyosis.

Pityriasis rubra, *dermatitis exfoliativa neonatorum*, or *Ritter's disease*, is a condition which, although not developed till extra-uterine life, has yet so many points in common with a mild case of attenuated foetal ichthyosis that I am not prepared to deny a close connexion between the two (*vide* description of this disease in Chap. XII.). Kaposi, in a letter published in the *Annales de Derm. et de Syph.* (3rd series, vol. iii. p. 453, 1892), states that there are times when "epidermitis exfoliativa neonatorum" can hardly be differentiated from ichthyosis, and suggests that it may be the same disease, beginning in the former case in extra-uterine life, in the latter within the uterus.

That *seborrhœa* is not the same as foetal ichthyosis may, I think, be concluded from the microscopic appearances found in the two conditions. The epidermic thickening in the latter condition is not dried sebum, but dried squamous epithelial cells, and the sebaceous glands are commonly atrophied, as was shown clearly by Caspary (16 and 17).

PROGNOSIS.

The attenuated form of foetal ichthyosis has not a grave significance as regards life. In five of the recorded cases (2, 3, 7, 24, and 29) death ensued ; but in three at least of these (2, 3, and 7) this could not be ascribed to the skin affection, for in No. 2 the child was also hydrocephalic, in No. 3 it had pernicious icterus and umbilical haemorrhage, and in No. 7 there was bronchopneumonia. The infant lived for twelve days in No. 3, for sixteen days in No. 2, and for eleven months in No. 7. In No. 24, in which death occurred on the third day, the cause may have been the ichthyosis, for no other lesion was found at the autopsy. Crocker's patient (29) had a low vitality from birth, and died in three months. All the other cases were alive when last seen by the observers, and had reached an age varying from seven days to sixty-one years.

With regard to the probability of complete cure there is not much hope. In one or two instances (4, 5, and 8), however, the child would seem to have recovered from the skin disease. In other instances there was localised involution of the malady (13, 14, 25, and 26), with, in Nos. 25 and 26, a tendency to revert. In most cases, however, the lesion either remained *in statu quo*, or showed an increase in severity as time went on.

TREATMENT.

Therapeutic measures have seldom been successful in this disease, a fact not to be wondered at when it is remembered how intractable is the adult form of the malady. The treatment adopted has been both internal and external, and the chief indications are to keep up the strength of the child by good feeding, nursing, and hygiene, and to soften and remove the epidermic covering.

The internal administration of medicines is of doubtful value. Seeligmann (1), finding signs of scrofula in his patient, administered antimony, mercury, and cod-liver oil; Weisse (12) gave alkalies internally; and G. H. Fox (13) prescribed the iodide of iron, with apparently some advantage. In most cases, however, the observer contented himself with keeping up the general health and strength by proper food, etc.

The external therapeutic measures generally took the form of warm baths, inunctions, and the use of lotions; and were for the purpose of removing the scales and keeping the skin moist and pliable. Simple warm (or alkaline) baths daily with soft soap inunctions have proved useful in certain cases, and ought to be tried. Friction with glycerine or with various oily substances (*e.g.*, cod-liver oil, lanolin, olive or almond oil, etc.) is of service both in removing the scales and in preventing their re-formation. Piffard (14) used local applications of peroxide of hydrogen, apparently with good results; and Behrend used unguentum diachyli. Salicylic acid, either in the form of a saturated solution in alcohol, or of a plaster, has been employed; and Perez (8) rubbed the whole body with an ointment containing tincture of iodine (9 grammes), glycerine (15 grammes), and cacao butter (15 grammes). Feeding with thyroid extract has not yet been tried, so far as I know, in this form of ichthyosis. It must be confessed that treatment has so far done little to mitigate the severity of the cutaneous lesions in this malady. Perfect cleanliness is, of course, a *sine quâ non*.

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ADDENDA.

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There is also in the *Anzeiger der K. K. Gesellschaft der Aerzte in Wien*, p. 203 (7th May 1885), a short note by Von Kogerer of a “Fall von Ichthyosis congenita mit Variola.” The child, a boy of seven years, had from birth been covered with a coat of grey-black scales. The head, forearms, hands, lower half of legs, feet, and genitals were free. He took variola, and there were plenty of pustules on the parts free from ichthyosis, a few near the margins of the patches, and none under the patches.

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CHAPTER IX.

CONGENITAL ICHTHYOSIS HYSTRIX.

MAJOR DEGREE: DEFINITION; SYNONYMS; HISTORICAL NOTE; AND CLINICAL HISTORY AND PATHOLOGY. MINOR DEGREE: DEFINITION; SYNONYMS; HISTORICAL NOTES; CLINICAL HISTORY; SYMPTOMATOLOGY; PATHOLOGY; PATHOGENESIS; DIAGNOSIS AND PROGNOSIS; TREATMENT. KERATOSIS PILARIS. LITERATURE.

IT is necessary to consider in this and in the following chapter certain cutaneous affections which are closely allied to one or the other of the two types of foetal ichthyosis which have been described in the preceding pages. Some of these diseases have been definitely recognised in the infant at birth, and may therefore justly be regarded as occasionally *fœtal*; others have not been noticed till a few days after birth, and can only be called *congenital* in the wide acceptation of the term. The first condition that falls to be discussed is ichthyosis hystrix. This disease occurs in two forms: in the one there is a wide-spread development of spines upon the surface of the body; in the other these formations occur only in limited areas, and can sometimes be recognised as following more or less closely the distribution of a cutaneous nerve or nerves. The former may be called the major degree, or marked type of ichthyosis hystrix; the latter the minor degree, or mild type of that disease. It may be at once remarked that all dermatologists are not in agreement about the validity of the claims of the second type to be considered as ichthyosis hystrix. It has by some been regarded as a variety of *Nævus*.

ICHTHYOSIS HYSTRIX (MAJOR DEGREE).

Definition.—Ichthyosis hystrix of the major degree may be defined as that variety of ichthyosis in which the epidermic thickening takes the form of wide-spread, but probably never

universal, papillary or wart-like growths, which vary in length, but often project to a distance of half an inch or more above the surface of the skin. Traces of this disease can usually be recognised at birth if carefully looked for, and it may even be that it has existed in a marked degree at that time. It is not usually fatal to life, but the lesion itself is very intractable.

Synonyms.—A semi-popular name for individuals affected with this variety of ichthyosis hystrix is “Porcupine Men,”—“Stachel-Schwein-Menschen” (German); “hommes porc-épics” (French),—and the malady itself has been called the “Porcupine Disease.” An occasional synonym is “ichthyosis cornea,” and another perhaps is “ichthyosis spinosa.” Other names have been given to it, as “ichthyosis cornea verrucosa” and “ichthyosis cornea acuminata” (Fuchs); “Hystricismus” and “Hystriciasis” (Plenk); and “Leontiasis hystrix” (Sauvage). Lebert gave to it the lengthy designation “keratosis diffusa epidermica extra-uterina.”

Historical Note.—A considerable number of cases of marked ichthyosis hystrix in childhood and adult life have been reported; but, with one exception, I shall deal only with those in which some morbid condition of the skin was recognised at birth. I make the exception in favour of the notorious Lambert family, in which the disease was present in a most marked form for several generations. The first member of this family was seen and described by John Machin in 1731 (“An Extract from the Minutes of the Royal Society, March 16, 1731, containing an uncommon Case of a Distempered Skin,” *Philosophical Transactions*, xxxvii. p. 299, 1733, Illustrated). The individual was then fourteen years old, was the only member of his family affected, and the condition of his skin had been compared to the bark of a tree, a seal’s skin, to the hide of an elephant or rhinoceros, to a great wart, or to the quills of a hedgehog. At birth his skin was normal, but in seven or eight weeks it became yellow, then black, and finally developed into the condition in which it was when seen by Machin. In 1755 H. Baker gave the further history of Lambert (“A Supplement to

the Account of a Distempered Skin," *Phil. Trans.*, xlix. Part I. for 1755, p. 21, 1756. Illustrated by G. Edwards). He was now 40 years of age and was still in the same condition. He had had six children all similarly affected, of whom one, a boy of eight, still survived. In all, the malady came on about nine weeks after birth. Both father and son had had small-pox, and during that time their warty growths had dropped off.* In the following year (1756) a description of the father and son appeared in French.† In 1802 Wilhelm Gottlieb Tilesius von Tilenau gave a full account of members of the third generation of the same family,‡ and many other writers gave more or less complete descriptions of them. A son of one of the two brothers seen by Tilesius was examined by W. Pickells in 1851 ("Cases of Ichthyosis and Hirsuties," *Edin. Med. and Surg. Journal*, lxxvi. p. 312, 1851); he was described in the handbills as "The Porcupine Man covered with scales, representing the skin of the rhinoceros," and it was stated that the morbid condition of the skin was first noticed when about two months old; he was now 35 years of age, and shed the covering of horny warts twice yearly. It is

* It is to these two individuals that the following advertisements from old London papers refer.

January 1755.—"To be seen at the Elephant and Castle, near Holborn Bars, the wonderful Production of Nature, and astonishing Porcupine Man and Youth his son, who are esteemed by all that have hitherto seen them to be the most extraordinary Production of human Nature, now or ever introduced to the Publick, and are at the present Time casting their Coats, to the Surprise of many Hundreds that daily resort to see them. Their solid Quills so numerous as not to be credited till seen."

Wednesday, February 26, 1755.—"To be seen at the George in Fenchurch Street a Man and his Son, that are cover'd from Head to Foot with solid Quills, except their Face, the Palms of their Hands, and Bottoms of their Feet. It's thought by the learned Physicians and many Hundreds that daily resort to see them, to be almost beyond the Power of Nature to produce two such Instances which are not to be credited till seen. Their Stay is but very short in this City."

"The Porcupine Man and his Son continue another Week at the George Inn in Fenchurch Street, by Reason there is a Subscription raised by many Gentlemen for him to be view'd on Friday next, at a Lecture-House in this City. Those that are desirous of seeing this wonderful Man and the fine Youth his Son who is no more than eight Years of age have still the Opportunity this Week and no longer."

† ASCANIUS—"Description d'un anglois d'une espèce singulière," etc. *Rec. périod. d'obs. de méd. de chir. et pharm.*, iv. p. 216. 1756.

‡ TILESIIUS (W.G.)—"Beschreibung und Abbildung der beiden sogenannten Stachelschwein-Menschen aus der bekannten Englischen Familie Lambert." Altenburg, 1802.

therefore evident that for at least four generations ichthyosis hystrix affected the males of the Lambert family, the first signs of the malady being noticed always about seven or eight weeks after the birth. It was not, therefore, strictly speaking, a congenital or foetal disease; but it afforded so striking an example of a hereditary morbid state the predisposition to which must have existed *in utero*, that its complete omission here could not have been justified.

With regard to ichthyosis cornea (s. hystrix) *fætalis* in the strict sense of the word, I have found only one case in medical literature which can with reason be regarded as a probable example of it, that, namely, reported by Ollivier (1)* in 1834. It must at the same time be remembered that in certain of the cases described as grave foetal ichthyosis (*vide* Smith's case in Chap. VI., p. 100 of this volume) small spines were found in certain regions of the body, but these were exceptional formations. Further, in one or two cases of ichthyosis hystrix developing during the first months of life it was noticed that the skin was not altogether healthy at birth; these were the instances reported by Duckworth (2), Crocker (3), and Sanders (4). It is probable that in the future careful inquiry into the early history of cases of ichthyosis hystrix may elicit details similar to those given in Duckworth's and Crocker's cases.

Clinical History and Pathology.—It will be most convenient to give separately the details of the cases described by Ollivier, Duckworth, Crocker, and Sanders, for the number of observations is so small that any analysis of them might prove misleading.

Ollivier (1) states that he was asked to examine the body of a foetus taken from the Seine. From the degree of putrefaction, he concluded that the infant might have been in the water for three weeks. It had the appearances of the full time, and there existed on the whole of the anterior aspect of the chest, and on the abdomen, especially in its upper half, an enormous number of warts (*verrues*) of a greyish-white colour, several of which had the size of a lentil. All were more or less manifestly pediculated, and the largest were split in the greatest part of their thickness. On detaching the epidermis that the putrefaction

* The figures in parentheses refer to the bibliography at the end of this Chapter.

had loosened from the skin, it was seen that all these excrescences traversed it without being covered by it. Dissection showed that each took root in the thickness of the derma as far down as the subcutaneous cellular tissue. It was not only on the chest and abdomen that these warty vegetations were seen ; they existed also on both shoulders, on the arms as far as the elbows, and especially on their outer and posterior aspects, and on the thighs and buttocks, but only externally and posteriorly. The regions named were covered to the same extent, on both sides of the body, by a considerable number of small warts, which gave a grained appearance to the surface of the skin ; several of them had the yellowish colour of ephelides, so that at a certain distance the skin seemed to be only altered in colour. All the organs of the foetus were in a healthy state. Ollivier, who regarded the growths as of the nature of warts, was much struck by their almost universal distribution, and by the fact that they were congenital. With regard to their pathological nature, he thought that they could not be venereal, for no analogy existed between them and the different kinds of syphilides ; but in the absence of any clinical history of the case, he was forced to leave the matter undecided. To my mind, it seems not improbable that this was really a case of foetal ichthyosis hystrix.

In Duckworth's patient (2), a boy thirteen years of age, with widespread ichthyosis hystrix and patches and streaks of xeroderma, it was noticed at birth that there were small red spots upon the body, and that in a few days heaping up occurred upon them.

Crocker (3) in 1879 described a weakly ichthyotic boy, and Sanders (4) saw him and noted his condition some years later. In this patient the mother noticed at the time of birth the appearance of bruising in three places on the thighs (the left thigh being the worst, and showing raw-looking areas as large as a finger). There were other abraded spots on the body and limbs. These healed up in about seven weeks ; but immediately warty growths began to appear about these places, and gradually spread over wider areas, until nearly the whole body was covered with them at the age of seven years, when the

process became stationary. The patches, which were described as raw at birth, were covered with thin cicatrices, except on the scalp, where they were like keloid. On the hands the growths were those of ichthyosis hystrix, whilst on the neck they were warty. The intervening areas of skin were healthy, not xerodermic. The mother had, in her pregnancy, suffered great hardships and privations. There was no suspicion of syphilis.

I shall not attempt to draw any conclusions from the study of these cases; but certain suggestions present themselves to my mind, and may here be stated. In the first place it seems probable, or at least possible, that the patients of Duckworth, Crocker, and Sanders may have carried *in utero* scattered horny projections, which being rubbed off during labour, gave rise to the bruised or raw-looking areas of skin. On the other hand, it must be admitted that possibly these abraded spots or patches may have been the seat of an intra-uterine pemphigus. For it will be remembered that according to Barkow (*vide* Chap. VII., p. 121) the first stage in the development of foetal ichthyosis is a pemphigus one. Further, in a patient with adult ichthyosis recently shown by Byrom Bramwell to the Medico-Chirurgical Society of Edinburgh (*Edin. Med. Journ.*, March 1894), and whom I was able, through Dr Bramwell's kindness, to interrogate, the fact was elicited that although the skin was normal on the first day of life, on the second it showed numerous blebs or blisters, and afterwards developed into the advanced state of ichthyosis in which it was found at the age of twenty-eight years. It is therefore very important that all the details of the commencement of this disease should be carefully inquired into.

In the second place the rarity of ichthyosis hystrix as a foetal condition is striking, and it would seem as if the intra-uterine state, whilst specially favourable to the production of the thick plates and scales of typical foetal ichthyosis, does not readily permit the formation of the spines and papillary projections which are the distinguishing feature in the hystrix variety.

With the exception of the two suggestions just made I do not think that the scope of this work justifies me in considering at greater length a disease (ichthyosis hystrix) which, although

evidently hereditarily predisposed to congenitally, has so rarely been noted at birth. It is, however, necessary to say a few words with regard to a morbid state which has been by some regarded as the *minor degree of ichthyosis hystrix congenita*, but which is generally designated *neuropathic papilloma*.

ICHTHYOSIS HYSTRIX (MINOR DEGREE).

Definition.—Ichthyosis hystrix of the minor degree, also called neuropathic papilloma of the skin, etc., may be defined as a skin disease usually present at birth, characterised by the presence of papillary growths, more or less pigmented, usually limited to one side of the body, and nearly always following the distribution of one or more of the cutaneous nerves.

Synonyms.—Many names have been given to this morbid condition, for some dermatologists have regarded it as a variety of nævus, others as a cutaneous neoplasm of a papillomatous nature, and others as a form of ichthyosis hystrix. It is the “papillary nævus” of A. T. Thomson, the “nævus neuroticus unius lateris” of Von Baerensprung, the “congenital papillary tumour” of F. Mason, the “neuropathic papilloma of the skin” of C. Gerhardt, Stephen Mackenzie, R. Pott, and G. E. Wherry, the “nerve-nævus” of Theodor Simon, the “papilloma neuroticum” of Allan Jamieson, the “nævus verrucosus unius lateris” of Brocq and Rivet, the “nævus verrucosus universalis” of Kaposi, the “ichthyosis linearis neuropathica” of Koren, the “ichthyosis cornea (hystrix) partialis” of Philippson, the “nævus linearis verrucosus” of Unna, and the “nævus linearis ichthyosiformis” of Lanz. Curtis called his case one of “ichthyosis hystrix congenita,” and Esmarch and Kulenkampff included nerve nævus and neuropathic papilloma among the varieties of “congenital elephantiasis.” Quite recently Hutchinson (38) has proposed for the condition the name “ichthyosis herpetiformis.”

Historical Note.—This skin affection has only been differentiated from others during recent years. Thomson’s two

cases (5) published in 1829, seem to be the earliest recorded instances of the malady, and Arndt (6) in 1839 noted another. Baerensprung in 1863 (7) first pointed out (in connexion with three cases) that the disease, like herpes zoster, had a nerve distribution, was limited to one side of the body, stopping exactly at the middle line. The case reported by H. Beigel under the name of "papilloma area-elevatum" (*Archiv für path. Anat. und Physiol.*, xlvi. p. 367, 1869) is now generally regarded as a bromide rash, and the patient seen by E. Geber ("Ueber eine seltene Form von Nævus der Autoren," *Vierteljahrsschrift für Derm. und Syph.*, N. F., i. p. 3, 1875) did not develope the disease till his fourteenth year. Mason (8) saw a case in 1871; and Gerhardt (9) noted two in the same year. T. Simon (10), under the name "Nerve-nævus" grouped two varieties: one, the trophic, in which the papillæ of the skin were hypertrophic; the other, the vaso-motor, in which there was enlargement of the vessels. O. Simon (11) observed a nerve-nævus in which, however, there were no papillary elevations, and Campana (12) put on record a series of twelve cases. Neumann's article (13), published in 1877, was an important one, and it was followed by useful contributions from the pens of Curtis (14), Hardaway (15), Stephen Mackenzie (16 and 23), and Radcliffe Crocker (17). Recklinghausen (18) considered this subject in his work on the multiple fibromata of the skin; and Esmarch and Kulenkampff (22) gave it a place near elephantiasis neuromatodes congenita in their large monograph on the forms of elephantiasis. Jamieson (19), Brocq and Rivet (20), Kaposi (21 and 28), and Cousland (22A), have also recorded cases which seem to have been congenital. In the children seen by Fox ("Papilloma of the Forearm," *Journ. of Cutan. and Venereal Diseases*, iv. p. 83, 1886) and by Robinson ("Nerve-nævus," *ibid.*, p. 147) the disease was not clearly congenital; whilst of the four cases reported by J. Jadassohn ("Beiträge zur Kenntniss der Nævi," *Vierteljahrsschrift für Derm. und Syph.*, xx. p. 926, 1888) only one, the fourth, seems to have been a genuine neuropathic nævus, and it was not noticed till the fourth day of life, although possibly congenital. I have, therefore, omitted the cases of Fox, Robinson, and Jadassohn from my bibliography (*vide* end

of Chapter). Pott (23A), Wherry (24), Hutchinson (25), Koren (26), Philippson (27), J. Müller (29), Little (30), Hagen (31), Barham (33), and Spietschka (36), have all reported congenital cases; but the patients seen by Petersen (32), Saalfeld (34), and Lanz (35), possibly did not come into the world with the cutaneous lesion fully evolved. Allusions to congenital neuro-pathic papillomata are also to be found in most of the recent text-books of Dermatology (e.g., Crocker's *Diseases of the Skin*, 2nd edit., pp. 347, 353) and in works on Pathology (e.g., Ziegler's *Path. Anat.*, English Trans., Part II., sect. i. to viii., p. 188, 1884). C. Kopp in his monograph (*Die Trophoneurosen der Haut*, Wien, 1886) fully discusses the pathogenesis of this interesting malady.

CLINICAL HISTORY.

The early history of most of the recorded cases of this variety of ichthyosis is very scanty, the chief reason being that they were not seen by the observer (usually a dermatologist) till some months or years after birth. In all the examples noted in the bibliography (*vide* end of Chapter), however, there was sufficient evidence to warrant the assertion that the skin was not quite normal when the infant was born.

A. *Maternal History*.—The mother would seem to have been usually a healthy woman with no skin disease of any kind; but details on this point are seldom given. In No. 14, however, the mother had a phthisical tendency. She was either a primipara (e.g., Nos. 13 and 16) or a multipara (e.g., Nos. 23 and 26). There was the history of a maternal impression during pregnancy in Nos. 29 and 34.

B. *Paternal History*.—The father also was usually a healthy man; but in No. 14 he suffered from subacute rheumatism, and in No. 16 from eczema and erythema of the hands.

C. *Family History*.—In No. 9α the father's brother had epilepsy and insanity (imbecility), and in No. 16 there was a family history of psoriasis. A sister of the patient in No. 23c had symmetrical morphœa, and an uncle in No. 14 had a congenital wart-like tumour on the neck. In the other cases the family history seems to have been free from skin disease, nervous

disorders, and syphilis. I have found no record in which more than one child in a family was affected with neuropathic papillomata.

SYMPTOMATOLOGY.

In all the cases in which any reference is made to the fact, the subject of this minor degree of ichthyosis hystrix was alive, well-nourished, and, save for the cutaneous lesion, well developed at the time of birth. The disease was pretty equally divided between the sexes, for of the thirty-eight cases in which information on this point was forthcoming twenty (Nos. 5 α and b, 9 b , 11, 14, 16, 17, 19, 20, 21 b , 23 α and b, 26, 29, 31, 33, 34, 36 α , b, and c) were males, and eighteen (Nos. 6, 7 α , 8, 13, 15, 21 α , 22 a , b, and c, 23 c and d, 24, 27 b , 30, 32, 35, 37, and 38) females. In a considerable number of cases, however, there is no record of the sex of the affected person.

The age of the patient at the time when he came under the observation of the reporter of the case varied very much. The presence of the skin disease was noted at birth or during the first days of life in Nos. 13 and 14; but in all the other cases the condition was not seen by the medical man till a later period, although it was observed by the mother or nurse when the child was born. In Nos. 23 c and 26 the child was nine months old when first examined by the observer; 1 $\frac{3}{4}$ years in No. 34; 2 $\frac{1}{2}$ years in No. 31; 4 years in Nos. 27 b and 28; 6 years in No. 9 α ; 9 years in No. 15; 10 years in No. 5 b ; 11 years in No. 30; 12 years in No. 19; 15 years in Nos. 17, 24, and 37; 16 years in No. 16; 18 years in Nos. 6 and 11; 19 years in No. 36 α ; 20 years in Nos. 8, 32, and 36 c ; 22 years in No. 23 d ; 24 years in No. 21 α ; 25 years in Nos. 5 α and 33; 28 years in No. 36 b ; 29 years in No. 20; 37 years in No. 29; 40 years in No. 21 b ; 47 years in No. 23 α ; and 61 years in No. 9 b .

With regard to the appearances, distribution, etc., of the cutaneous lesion at the time of birth, it may be well here to state briefly the descriptions given by Neumann (13) and Curtis (14). In Neumann's patient, a well-developed female infant, there were found on the right nates and right lower limb prominent dull-white outgrowths arranged in regular lines and bands.

These on the soles of the feet had the appearance of blisters, but elsewhere they evidently consisted of firm, pointed and flat papillary hypertrophies of about the size of a millet seed. On the nates and on the outer surface of the right thigh these were arranged in bands from 1 to 2 cms. in breadth ; on the perineum and around the labia majora were circular bands about 3 mms. broad ; and on the outside of the right leg were three narrow streaks about 1 cm. in length by 2 mms. in breadth. On the dorsum of the right foot were three parallel stripes, each 5 mms. in breadth ; one passed from the heel to the little toe, another went to the second toe, and another to the fourth. On the sole of the foot were two streaks 5 mms. in breadth, beginning at the heel and extending to the little and fourth toes respectively. The skin lesion was found situated exactly in the areas of distribution of certain of the cutaneous nerves of the parts. During the first two months of life the growths increased somewhat in size, and those on the soles became yellow in colour ; but thereafter involution began, and they disappeared first in the region of the calf, then in that of the thigh, and finally from the feet.

In Curtis's patient (14), a vigorous and otherwise healthy male infant, the eruption was seen at birth, but was supposed to be due to an accumulation of vernix caseosa. When the child was a week old the lesion was seen to consist of papules or papulo-tubercles, each about half the size of a split-pea, circular in outline, and slightly elevated, and having a uniformly brownish colour. These were arranged in irregular groups of various forms, with fairly well-defined margins. The surface was slightly scaly and had a rough feeling ; but the scales were adherent, and there was no desquamation. The lesion was confined to one side of the body (the left), and affected chiefly the arm and leg, spreading a little on to the chest, but only in the form of scattered, less well-developed spots. It was best marked on the knee, where on the inner and anterior aspect was a somewhat circular patch about two-thirds the size of the adult palm, of a brownish colour, quite dry, showing no cracks, and exhibiting only the slight depressions between contiguous papules. The papules were well-marked, and the patch was visibly but only slightly scaly. An elongated patch stretched downwards along

the front of the leg and dorsum of the foot to the extremity of the hallux, the nail of which was not affected. This streak was made up chiefly of distinct papules, confluent only in limited areas; and there were some outlying papules near it. Starting again from the great toe, a small group was found on the plantar aspect of the foot, and from this was another linear group which ran along the sole. This rounded the inner ankle and proceeded up the calf of the leg to the popliteal space, where it joined the large patch already described. This streak was made up almost entirely of a single-file arrangement of papules without outlying formations of any kind. From the patch at the knee the lesion extended up the inner and anterior aspect of the thigh in the form of scattered papules; this ended at the groin and did not spread to the genitals or trunk. On the palmar surface of the fingers and hand were scattered or slightly grouped papules, which were most marked on the ulnar aspect. These extended up the inner side of the arm to the shoulder in an irregular fashion, and the papules passing along the anterior edge of the axilla, without invading that space, radiated downwards over the anterior and inner aspects of the chest. The patches on the upper part of the body were thinner and of a lighter colour than those on the leg.

I have given the details of these cases somewhat fully, for they were fairly typical ones, and were observed by the medical man at, or soon after, the birth of the affected infant. It will be necessary, however, to mention some of the characters of the eruption in other cases.

With regard to distribution, the lesion was almost invariably strictly unilateral; but in No. 19 it was bilateral, and in one or two other instances a few papules extended slightly across the middle line. In the great majority of cases it was accurately limited by the middle line of the body (*e.g.* mid line of sternum in front, spines of vertebrae behind). It was right-sided in Nos. 5 α and b ; 9 α ; 11; 12 e , f , i , and k ; 13; 16; 20; 23 b and c ; 24; 26; 27 a and b ; 29; 30; 32; and 33. It was confined to the left side of the patient in Nos. 6; 7 α and c ; 8; 10; 12 a , b , c , and g ; 14; 15; 17; 23 a and d ; 31; 35; and 36 a and c . In No. 9 b it was right-sided, save for a small patch in the first intercostal space; and in Nos. 12 d , h , and l , 19, and 36 b , it affected

both sides. In No. 37 the lesion was almost entirely to the right of the middle line, there being only a few patches on the left side.

The distribution areas of the cutaneous lesion are given in the accompanying table, and it will be seen from it that any part of the body may be affected. In a few instances the lesion had a universal distribution ; but in most cases it was more or less local. In nearly every patient it could be definitely traced that the wart-like projections followed the lines of certain of the cutaneous nerves of the part.

TOPOGRAPHY OF ICHTHYOSIS HYSTRIX (MINOR DEGREE).

No. of Case.	Area of Distribution.
5a.	Nipple, axilla, inner side of arm, palm of hand, little and ring fingers.
5b.	Lip and chin.
6.	Arm, neck, thorax, and abdomen.
7a.	Neck, shoulder (from outer end of clavicle to mid line of sternum), upper arm, and lower part of chest. Affected especially the area of the 4th cervical nerve.
7b.	Area supplied by 3rd cervical nerve.
7c.	Area supplied by 5th dorsal nerve.
8.	Abdominal wall below and to left of navel.
9a.	Thorax, abdomen, upper arm, and radial side of forearm (right side) ; and left side of nose, upper lip, left cheek, and behind left ear.
9b.	Axilla, upper arm, elbow joint (inner aspect), thigh, penis, and scrotum (right side), and first left intercostal space.
10.	Cheek and upper lip (left side), with congestion of mucous membrane of hard palate, tonsil, and pharynx.
11.	Back of shoulder and upper arm, especially in area of posterior branches of the lower cervical nerves.
12a.	Area of 1st and 2nd branches of trigeminus.
12b.	Area of 2nd branch of trigeminus.
12c.	Area of 2nd and 3rd branches of trigeminus.
12d.	Area of 1st and 2nd branches of trigeminus (both sides).
12e.	Area of cutaneous branches of 3rd and 4th cervical nerves and of 3rd branch of trigeminus.

No. of Case.	Area of Distribution.
12 <i>f.</i>	Area of brachial plexus.
12 <i>g.</i>	Area of one anterior intercostal nerve branch.
12 <i>h.</i>	Area of posterior branches of spinal nerves (both sides).
12 <i>i.</i>	Area of cutaneous branches of 1st lumbar nerve.
12 <i>j.</i>	Area of cutaneous branches of pudic nerve.
12 <i>k.</i>	Area of crural nerve (cutaneous branches).
12 <i>l.</i>	Area of cutaneous branches of sacral plexus (both sides).
13.	Nates, perineum, lower limb (outer side), and both dorsum and sole of foot.
14.	Leg, arm, and slightly on thorax.
15.	Arm, leg, and trunk (face and neck exempt).
16.	Penis, scrotum, perineum, nates, inner and anterior aspect of lower limb down to knee.
17.	Arm, leg, neck, shoulder, and in line of ribs.
19.	Neck (both sides).
20.	Scalp, face, neck, thorax, abdomen, and limbs.
21 <i>a</i> and <i>b.</i>	} Universal distribution.
22 <i>a.</i>	Scalp, face, neck (right side), and on left side of thorax, upper limb and lower limb (thigh).
23 <i>a.</i>	Trunk, groin, scrotum, upper and lower limbs.
23 <i>b.</i>	Shoulder and chest.
23 <i>c.</i>	Anterior aspect of chest and inner side of upper limb.
23 <i>d.</i>	Neck (area of cervical plexus).
24.	Upper limb (hand and wrist), lower limb, and abdomen.
25.	Back and lower limb.
26.	Upper limb.
27 <i>a.</i>	Trunk and lower limb.
27 <i>b.</i>	Lower limb.
28.	Especially on the limbs.
29.	Neck, ear, axilla, scrotum, and thigh.
30.	Whole of right side of body.
31.	Neck, arm, and thigh.
32.	Neck, chest, thigh, and leg.
33.	Check, hand, back, scrotum, thigh, and hallux.
34.	Neck, chest, arms.

No. of Case.	Area of Distribution.
36a.	Arm and chest.
36b.	Arms and trunk.
36c.	Back, abdomen, and thigh.
37.	Axilla, thorax, abdomen, and buttocks.

An idea of the appearance of the cutaneous lesion will have been obtained from the description given in the two cases (Nos. 13 and 14) noted above; but certain general facts may here be stated. The disease consists in the presence of papillomatous or warty growths, sometimes arranged in patches of the size of a crown piece or smaller, and at other times running in streaks or lines following the distribution of certain of the cutaneous nerves. They are often more or less deeply pigmented, and the surrounding skin is also sometimes the seat of pigmentary deposits. The papillomata themselves vary considerably in size, and have been compared to a pin-head, a pea, or a millet seed. They are either soft or moderately firm in consistence. Sometimes they are closely aggregated, at other times they are widely scattered. A scanty, but usually an offensive, secretion is sometimes present, and occasionally the presence of itching is noted. In many cases, although the lesion is undoubtedly present at birth, it becomes more evident (through growth in size or pigmentation) as age advances.

The general health of the patient does not seem to suffer much, if at all, from the presence of these warty growths on the skin. It is interesting, however, to note that in certain cases the cutaneous lesion occurred in individuals suffering from nervous complaints. For instance, Gerhardt's first patient (No. 9a), a child of six, had epileptic fits which commenced about the third year, and showed some weakness of intellect; whilst in the case reported by Radcliffe Crocker (No. 17), a boy of fifteen, there was pseudo-hypertrophic paralysis. In many instances, however, as in No. 24, there was neither a neurotic history nor the presence of any nervous disease. One of Mackenzie's patients (16) had paraphymosis, and sometimes other skin diseases, such as pigmentary nævi or elephantiasis, were present as complica-

tions. There was club-foot in No. 31, and the boy was poorly developed in both mind and body. Arndt's patient (6), although eighteen years of age, had never menstruated, and had not even had menstrual molimina ; there were no hairs on the mons veneris, but the mammae were developed (*vide* Plates VII. and VIII.).

PATHOLOGY.

The naked-eye appearances of the affection have already been described, and in a few words I may indicate the microscopic characters which have been found. Gerhardt (9), for instance, observed that the lesion was a tumour-growth affecting the cutis and the epidermis. The former showed great richness in its lymph cells, and its connective tissue was loose in structure ; the arterial twigs were proportionately large. Towards the surface the cutis was projected into knobbed or finger-shaped polypi, from .5 to 1 mm. in length. These contained vessels, and consisted of fibrillar connective tissue, here and there rich in cells. These papillæ were covered with cylindrical cells which resembled those of the lowest layer of the rete Malpighii, but differed from them in possessing much brownish-yellow pigment. On these lay flat cells with hyaline contents, and there was a thin layer of horny cells on the surface. Wherry (24) found that an excised piece of affected skin contained small papillomata with a thickened epidermis ; there was no cellular infiltration of the cutis, and no obvious change in the nerve branches ; there was slightly increased vascularity of the enlarged papillæ. Philippson (27) also gives an account of the microscopical appearances of the affected skin. T. Simon (10), it may be noted, divides nerve nævi into two varieties, trophic and vasomotor ; in the former the papillæ are hypertrophied and the rete Malpighii pigmented, whilst in the latter there is increased formation and widening of the vessels.

The general statement may be made that the pathology of this disease is sometimes that of ichthyosis hystrix and sometimes that of pigmented soft warts.

PATHOGENESIS.

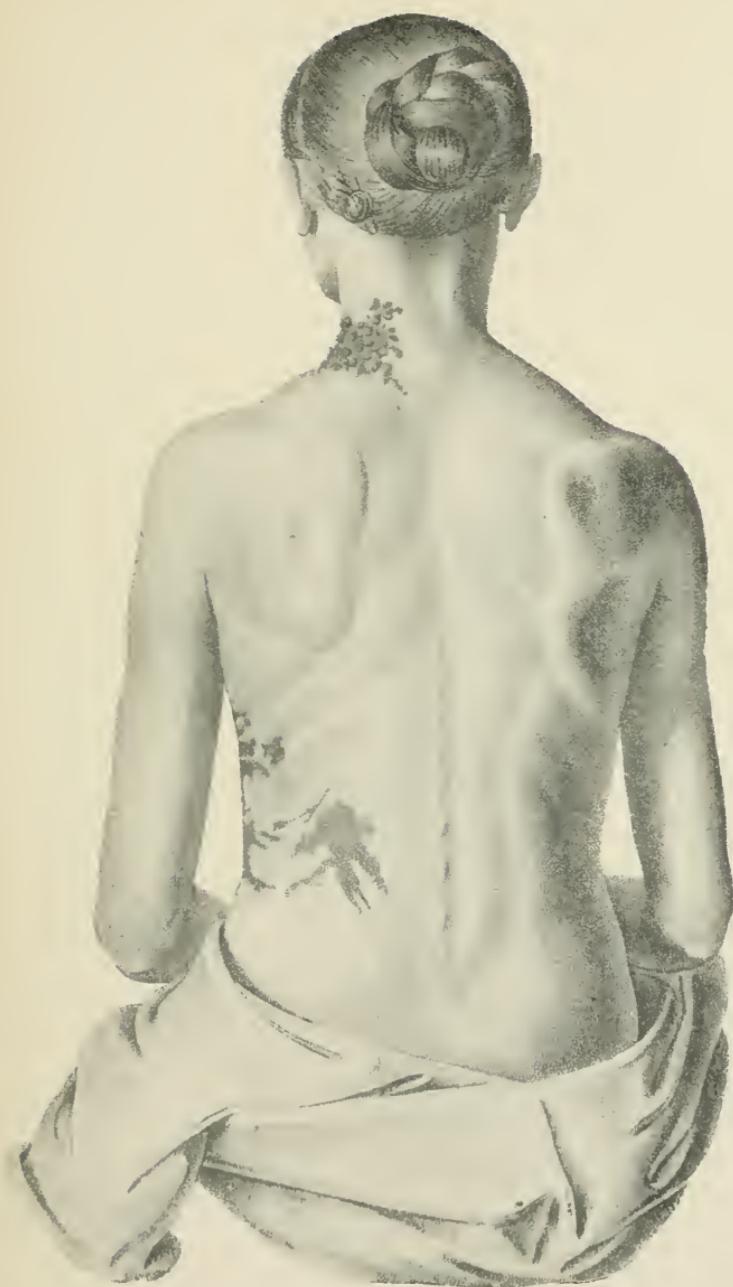
The fact that this congenital skin affection has been called by

PLATE VII.



ARNDT'S CASE OF NERVE-NÆVUS.

PLATE VIII.



ARNDT'S CASE OF NERVE-NÆVUS.

so many different names may be taken as proof that various opinions are held as regards its *nature*. These opinions may roughly be arranged in three groups.

(1.) Many dermatologists have regarded the disease as a variety of nævus, and have in consequence applied to it such names as nerve nævus, nævus verrucosus, papillary nævus, and nævus verrucosus unius lateris—names which convey the idea of its possible nervous origin, of its warty or papillary nature, and of its unilateral distribution (Nos. 5, 6, 7, 10, and 20).

(2.) Other writers have considered that it is most closely allied to ichthyosis, and have considered it to be a mild form of ichthyosis hystrix of congenital origin (Nos. 13, 19, and 26). Ziegler, for instance, thinks that it is closely allied to, if not identical with, ichthyosis.

(3.) Others, again, have thought it best to look upon it as a separate disease, and not as a form of nævus or of ichthyosis, and have suggested such names for it as neuropathic papillomata, neuropathic warts, papillary growths of nervous distribution, and papillary neuromata ("neurome papillaire" of Labb  and Legros).

Hutchinson would seem to regard it as the intra-uterine variety of herpes zoster, and other writers have been struck by the resemblance in its distribution which it bears to that disease.* The theory that it is a syphilitic skin disease—a maculo-papular syphilide—seems scarcely to have been advanced at all, probably on account of the absence of a syphilitic history in any of the recorded cases. Whilst recognising its affinity with ichthyosis hystrix, congenital warts, and warty forms of nævus, I am personally inclined to regard it, not so much as a separate disease, but rather as an intermediate type or connecting link between the above-named morbid states. Indeed, there can hardly be said to exist any clear line of demarcation between an ichthyosis hystrix which occurs in the form of scattered papillary projections and congenital verruc  or warty nævi.

* In a recent paper, Hutchinson (38) says that its deviation from close similarity to herpes zoster may be explained by the fact that the disturbance of the nutrition of the skin occurs at an early period of foetal life, before the nerve structures have become developed according to their final plan.

With regard to the *mode of origin* of the disease, a nervous theory seems to be that which has been most commonly suggested. Thus Hutchinson thought that it might be caused by intra-uterine herpes zoster, and von Baerensprung (7) considered it as due to disease of the spinal nerve ganglia in the fœtus. T. Simon (10), also, ascribed it to a disturbance of vaso motor and perhaps of trophic nerve branches in foetal life, and divided it into two varieties according as the former or the latter were affected. Campana (12) adopted this theory of origin, and Neumann (13) suggested that the extra-uterine increase in the extent of the lesion might be due to further development of the intra-uterine nerve disease. Stephen Mackenzie (23) regards it as probable that "some irritation or disturbance of the central or trophic nervous system occurs at a time of developmental activity in the skin previous to birth, and leads to some perversion of its development, overgrowth of the papillæ or connective tissue, and disarrangement of its pigmentation." It must, however, be remembered that such nervous lesions are purely hypothetical ; they have never been demonstrated.*

DIAGNOSIS AND PROGNOSIS.

The congenital nature of the disease, its unilateral distribution, and the fact that the wart-like projections are usually limited to the area of certain cutaneous nerves, will generally serve to differentiate this malady from other morbid states ; but, as has been stated already, it cannot always be clearly marked off from certain forms of nævus or of congenital warts. The lesion is not in itself dangerous to life, although it may give rise to considerable irritation, and occasionally, when located in the face, to disfigurement. Occasionally it has a tendency to spontaneous involution (*e.g.*, No. 13) ; but much more commonly it increases in extent and in darkness of pigmentation as the patient grows older.

* Kopp (*Die Trophoneurosen der Haut*, 1886) denies that it depends upon an intra-uterine affection of the spinal ganglia, and ascribes it to a congenital abnormal arrangement of the smaller bloodvessels along the course of the peripheral branches of certain nerves.

TREATMENT.

Treatment has usually consisted in the destruction or removal of the wart-like growths ; thus Thomson (5) destroyed them one by one by means of strong nitric acid, or removed them by the knife, and Mason (8) also performed excision of a good-sized patch of papillomata. The irritation produced by the growths may be allayed but not removed by the use of salicylic acid plaster, etc. Wherry (24) tried the effect of two methods of treatment on his patient : on one part of the body he destroyed the papules (under ether) with potassa fusa, but for the malady as it appeared near the wrist he cut down upon the musculo-spiral nerve where it emerged from under the supinator longus, and divided it. The result was better with the latter method, for in six months there was only a slight roughness left on the wrist, whilst on the other parts of the body the scars itched as badly as ever. Crocker (37) has tried the effect of thyroid feeding, but with only temporary benefit.

KERATOSIS PILARIS.

A few words may here be said with regard to a disease which has not yet been actually recognised at the time of birth, but which is met with almost constantly in the young, and has certain affinities with ichthyosis. Like many other diseases belonging to the group of the keratoses, it has been described under several different names. It was the "folliculitis rubra" of Erasmus Wilson, and the "cacotrophia follicularum" of Tilbury Fox. Neumann, Duhring, and others called it "lichen pilaris," a name best reserved, according to Crocker, for the *inflammatory* disease of the hair follicles ; and Auspitz and Lesser have named it "ichthyosis follicularis." Other designations have been "follicular xeroderma" (Liveing), "pityriasis rubra pilaris" (Duhring), "keratosis" or "hyperkeratosis follicularis" (Kaposi), "ichthyose ansérine des scrofuleux" (Lemoine), "ichthyose cornée" (Hardy), "pityriasis pilaris" (M'Call Anderson), "xérodermie pilaire" or "ichthyose anserine juvenile" (Thibierge), "xérodermie pilaire érythémateuse" or "congestive progressive" or "ichthyose rouge"

(Besnier), and "ulerythema ophryogenes" (Taenzer). Perhaps, however, the most commonly used and the best name has been "keratosis pilaris."

The various terms mentioned above reveal much with regard to the nature and characters of the disease. They tell us that it is a lesion of the hair follicles, and that the nature of the lesion is a keratosis,—an accumulation of the cells of the stratum corneum which in this instance plug the orifice of the follicle and form a small papule (lichen). They tell us also that it is related closely to ichthyosis and to its mildest manifestation, xeroderma; that it resembles pityriasis and that it might be mistaken for cutis anserina (save that the latter is a transitory condition); that it is characterised by redness of the integument; and that it occurs commonly in the young and scrofulous.

A few other facts may be noted in addition to those given above. The papules, which are of the size of a pin's head, lie at the orifice of a hair follicle, and are sometimes pierced by the hair itself, are found either on the face or on the trunk and limbs (especially on their extensor surfaces). The papules vary in colour from white to rosy red, and from red to brownish black, and the intermediate skin is coloured like the papules themselves. The adjacent integument, also, may be xerodermatosus and even ichthyotic, and to the touch the whole lesion has a "nutmeg grater" feeling. The hairs are almost always in an atrophic condition, and may, in some instances, have quite disappeared. Subjective symptoms may be quite absent, but sometimes there is a certain degree of irritation, leading to scratching.

The microscopical characters of the lesion have not yet been sufficiently worked out; but, according to Lemoine,* there is a hypertrophy of the derma and atrophy of the hairs and sebaceous glands, the hair follicles and sudoriparous glands are surrounded by numerous embryonic cells, the subcutaneous adipose tissue shows inflammatory changes, the cells of the deeper layers of the epidermis and those of the rete Malpighii are elongated vertically, the stratum granulosum is little marked, and the cells of the stratum corneum are enlarged individually and are arranged in a great number of layers.

* Lemoine, *Annales de Derm. et de Syph.*, 2nd ser., iii. pp. 274 and 343, 1882.

The lesion is said to begin to be manifest about the second or third year of life, and sometimes later; but in some cases it "had always been present," and in one instance that I have seen it was probably present soon after, if not actually at the time, of birth. The facts that it is sometimes hereditary (the mother having or having had the same disease as her infants), and that it often shows "family prevalence," taken in conjunction with the circumstance that it is certainly a keratosis, and very probably an attenuated form of ichthyosis, lead one to believe that it will yet be recognised by some careful observer at the time of birth. It should also be noted that it occurs in the descendants of individuals affected with undoubted ichthyosis, and that it is often complicated by a xerodermatos condition of the skin elsewhere in the same patient. Probably it is at least as truly "congenital" as most cases of ordinary ichthyosis.

The treatment that has been usually adopted, although not with great success, has been the internal administration of cod-liver oil and arseniate of soda, and the external application of glycerine soap, carbolic acid ointment, and plasters of cod-liver oil, salicylic acid, resorcin, etc.

The disease has been very fully described by L. Brocq, to whose admirable monograph the reader is referred for a good account of the history of the malady and of its manifestations.* It may here be noted that the case described by J. C. White under the name of "keratosis (ichthyosis) follicularis" (*Journ. of Cutaneous and Genito-Urinary Diseases*, vii. p. 201, 1889) is, according to Brocq, really an example of Darier's disease (psoriasis folliculaire végétante). Brocq's paper, above noted, contains also the references to the works of Erasmus Wilson, Tilbury Fox, Neumann, Duhring, Kaposi, Lemoine, Auspitz, Liveing, Hardy, Lesser, Jackson, Hyde, M'Call Anderson, Thibierge, Besnier, and Taenzer.

* Brocq (L.), "Notes pour servir à l'histoire de la Keratose pilaire," *Annales de Derm. et de Syph.*, 3rd ser., i. pp. 25, 97, and 222, 1890.

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CHAPTER X.

DISEASES OF THE SKIN (Continued).

TYLOSIS PALMÆ ET PLANTÆ: DEFINITION; SYNONYMS; HISTORICAL NOTE; CLINICAL HISTORY AND SYMPTOMATOLOGY; PATHOLOGY; PATHOGENESIS; TREATMENT. AKROKERATOMA HEREDITARIUM (NEUBURGER). TYLOMA. CLAVUS. CORNU CUTANEUM. LITERATURE.

THE forms of congenital ichthyosis which I have considered in the preceding chapters belong to the family or order of the "keratoses,"* and there remain one or two diseases, which are sometimes present at birth either actually or potentially, which must be grouped with them. They are all "hyperkeratoses," *i.e.*, conditions in which there is an excessive development of the horny layer of the epidermis, and may be considered in the following order: Tylosis palmæ et plantæ, callositas, clavus, and cornu cutaneum.

TYLOSIS PALMÆ ET PLANTÆ.

Definition.—The disease which has been named tylosis palmæ et plantæ may be defined as a hypertrophy of the horny layer of the epidermis affecting only the palms of the hands and the soles of the feet, and leading to the development of a hard plate in those regions.† It is occasionally an acquired condition, but is more commonly congenital in, at any rate, the wide sense of the word. It is to be distinguished from the secondary and localised thickening in some cases of psoriasis, eczema, syphilis, etc.

Synonyms.—The following terms may be regarded as synonymous with tylosis palmæ et plantæ (Crocker): "Ichthyosis palmaris et plantaris" (Thost and others); "tylosis palmæ manus plana" (Hebra); "keratoma plantare et palmare hereditarium"

* Auspitz (H.), *System der Hautkrankheiten*, p. 119, 1881.

† Crocker (H. R.), *Diseases of the Skin*, 2nd edit., p. 363, 1893.

(P. G. Unna); and "inherited keratosis of the palms and soles" (Hutchinson, G. H. Fox, and others). H. Hebra simply regards it as a variety of "tylosis" or "callositas," without giving to it any special designation.*

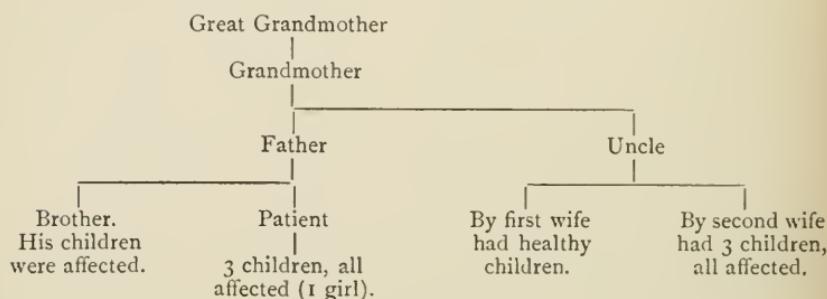
Historical Notes.—According to Unna (7), Ernst (1) and G. Simon (*Die Hautkrankheiten*, p. 48, 1851) were the first to record cases of this disease. In 1851, also, Pickells (2) noted what he regarded as a congenital ("connate") instance of it in a boy of 13 or 14 years of age, and in 1879 Bulkley (3) saw a woman of 25 in whom it had been present "since infancy." Thost's inaugural dissertation (4) contained a most interesting account of a family in which this disease was hereditary. Other cases were those noted by Boegehold (5), and Startin (6); and P. G. Unna (7) made a most important contribution to our knowledge of the malady in 1883. Since that year G. H. Fox (8), C. M. G. Biart (9), W. H. Date (10), Jonathan Hutchinson (11, 14, 15, and 16), H. R. Crocker (12), and H. G. Brooke (13), have all written upon the subject, and have added to the list of known cases.

CLINICAL HISTORY AND SYMPTOMATOLOGY.

The most striking fact in the history of cases of keratosis palmæ et plantæ is *heredity*. This heredity manifested itself either in the existence of the disease in parent and child, or in the occurrence of "family prevalence," or in both these forms at the same time. In No. 2 the other children of the family were affected in the same way as the patient; and in No. 3 the father, the father's brother, and the father's father all had it. Thost's record (4) was a most remarkable one, for there were seventeen cases in four generations of the same family, eight of which were seen by the author himself: the great-grandfather of the little girl (Lina Neubrandt) had keratosis palmæ et plantæ; he had seven children, six males and one female, of whom five (all males) inherited the disease; of the five affected males, one died young, one was lost sight of, two had families in which there was at least one case of the malady, and the fifth (Heinrich Heilmann) was twice married, and had a large family in which

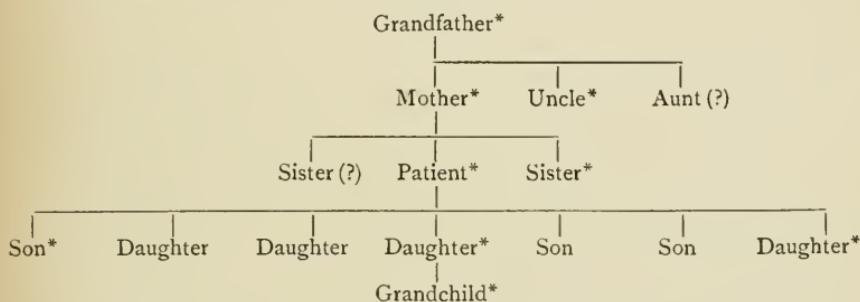
* Hebra (H. von), *Die Krankhaften Veränderungen der Haut.*, p. 361, 1884.

the cutaneous anomaly was very evident ; Heinrich Heilmann had eight children, five males and three females, of whom six (four males and two females) were similarly affected ; finally, the eldest daughter of Heinrich (Margarethe Neubrandt) had four children, three girls and one boy, and of these, all the girls, including Lina Neubrandt, the first of this remarkable family who was seen by Thost, had it. A more complete demonstration of heredity, both the direct form and that known as "family prevalence," could scarcely be desired. In Boegehold's case (5), on the contrary, heredity was apparently absent, and in No. 6 no information on the point is forthcoming. Unna's two observations (Nos. 7a and b), however, again clearly point to heredity : in No. 7a there were eight cases (five males and three females) in three generations ; whilst in No. 7b (the Ebert family) the mother had it as well as three of her four children (two males and one female), the fourth infant dying when three months old, and showing no evident signs of it at the time of death. The first patient seen by G. H. Fox (No. 8a) was one of eight children, every second one of whom inherited the disease, and his mother, mother's mother, an uncle, and two nephews were similarly affected ; whilst the father of Fox's second case (No. 8b) had also the disorder. In No. 9 the patient's father, his father's brother, and all his brothers and sisters, had keratosis of the palms and soles. The list of affected individuals in connexion with W. Horton Date's patient (No. 10) may be represented in a tabular form.



Hutchinson has seen two cases of the disease: in the first (No. 11) only the patient himself and his three sisters had keratosis ; and in the second (No. 14) three of the patient's

sisters had xeroderma, and his father had "hard hands," but none of his children nor his sister's children were affected. Hutchinson also has resuscitated a case (No. 15) of what he believes to be, in its early stages at least, an instance of keratosis of the palms and soles,—that reported by W. Sömmerring in the early part of the century; in it there is unfortunately no record of the family history. Brooke (No. 13) has seen two cases, in both of which the disease was present in the mother, in her parents, and in one or more of her children. In conclusion, I may give the family tree of Crocker's patient (No. 12).



In this case, which closes this remarkable demonstration of heredity, nine individuals in five generations were certainly affected with the disease, four were not diseased, and with regard to two there was some doubt.

There is no limitation in the matter of *sex*, for an analysis of the preceding family histories shows that fifty-one males and thirty-one females were affected. The more active habits of the male may possibly account for the fact that men seem to have developed the disease more often than women.

With regard to the question, at what *age* the cutaneous lesion was first recognised, all the writers who have described the cases enumerated agree in stating that it appeared in early infancy. Some affirm, but on hearsay evidence, that it was present at birth, and others note that it first became evident when the infant began to crawl about. In all Thost's cases (4) it was said that the disease was observed in the first week of life as a slight roughness of the epidermis of the palms and soles, which rapidly increased in thickness. With regard to the second family seen by P. G. Unna (7), the interesting remark is made,

that immediately after birth both palms and soles were observed to be surrounded by a bluish border, on which, after the seventh month of life, horny growths began to appear, and from which these growths soon spread to the rest of the palmar and plantar surfaces. It may be concluded that if the skin of these parts were carefully examined at birth some slight morbid change would be discovered in some cases at least. At any rate, the predisposition to the disease is congenital, although pressure and friction may be necessary to develop the characteristic lesion.

In all the recorded cases both the palms and soles were affected, and it is from this typical *distribution* of the lesion that the disease has got its name. In Nos. 2 and 5, however, a condition similar to that on the palms and soles was found on the knees, and in Nos. 11 and 14 there was general xeroderma. No. 8b was an interesting case, for in it there was keratosis pilaris of the arms and thighs, as well as general ichthyosis of a mild type, and tylosis palmæ et plantæ.

With regard to the *characters* of the lesion itself, one or two of the descriptions given by authors may here be stated. Startin (6) described it as consisting of dense, yellowish-brown plates of epidermis on the plantar surface of the feet and on the palms of the hands, having well-defined margins. The skin appeared to be harsh and dry, was wrinkled and very stiff, and had little or no fat in its substance. Of the two cases reported by Crocker (12), the first was an acquired example, but the second was congenital. In the latter the whole surface of the palms and soles was covered by a uniformly thick plate of horny epidermis of a yellowish-white colour, just like a layer of wax ; this terminated abruptly at the sides of the hands and fingers. The main lines of the hand were deepened, but the smaller ones were absent. There was also marked thickening of the horny layer on the extensor aspect of the joints of the fingers. With the exception of the inner border of the arch of the foot, the soles were similarly affected ; but the epidermis was thicker and had a greater tendency to split. The part of the sole affected was that which touches the ground when the patient stands ; but on the clear areas were some corn-like formations. The condition had existed since birth ; but every autumn blisters formed

beneath the epidermis, and the whole horny layer peeled off in about two months. In some cases itching was a prominent symptom ; and although, as a general rule, there was xeroderma, yet in No. 7 there was osmidrosis and hyperidrosis of the soles, showing that the keratosis did not block up the canals of the sweat-glands. In No. 7, also, the sensibility to touch was unimpaired ; but there is commonly diminished sensitiveness. In No. 5 the nails were curved on themselves and ragged.

The general health of the patients seems always to have been good, although in No. 3 there was rheumatism.

PATHOLOGY.

The horny plate on the palms and soles had a thickness varying, in most cases, from one-eighth to one-sixteenth of an inch, and its surface was either smooth or pitted. The other macroscopic characters of the lesion have been already mentioned. With regard to the microscopic appearances of the diseased integument, Thost (4) gives a full description, which may be thus summarised :—The papillæ were increased in length fivefold, their breadth was somewhat less than normal, and their number was not greater than usual. The prickle-cells were not enlarged or otherwise altered ; but their number was greatly increased, and on this account the rete Malpighii was much broader. The stratum granulosum was normal. The horny layer was much thicker than in the healthy condition, and the increase was chiefly in its middle layer. The cutis, as a whole, was thicker than usual, and the adipose tissue more marked. The bloodvessels appeared to be enlarged.

PATHOGENESIS.

The names which have been given to this disease by various writers indicate that all are not agreed as to the *nature* of the morbid process. Some have considered it as a localised form of ichthyosis ; but others have regarded it as more closely allied to cutaneous horns Unna (7), for example, places it in the group of the acanthoses along with verruca, condyloma acuminatum, cornu cutaneum, tyloma, and clavus ; and, indeed, looks upon it as a *variety* of cornu cutaneum, giving ten reasons why it should

be regarded as more nearly allied to that disease than to ichthyosis. Bronson* adopts Unna's view of the matter. The decision with regard to its exact place in any system of classification must be left to skilled dermatologists ; but to my mind its resemblance to the grave type of foetal ichthyosis is very striking. There is certainly marked hyperkeratosis, but whether or not there is also hyperacanthosis must be settled by future microscopic research.

With regard to its *mode of origin*, opinions differ. It is probable that even in the cases in which the malady did not appear till late in life there was a congenital predisposition to it. It would seem, however, that the predisposition is rarely strong enough to bring about the full evolution of the disease, and that an exciting factor, in the form of pressure or some other irritant, is necessary to develope the characteristic lesion. This view is supported by the markedly hereditary nature of the malady, which has been already referred to. Hutchinson (16) says, with regard to one of his cases, a girl of five: "No doubt the child was born with a peculiarity in the structure of the skin which rendered it less capable than usual of resisting the effects of mechanical irritation ; thus, when the child began to walk and to handle toys the skin revealed its defective organisation by passing into a condition of chronic inflammation. In this respect we have a parallel with certain other well-known types of disease, such as retinitis pigmentosa, Kaposi's disease, and many others in which nothing is to be observed at the time of birth, but in which exposure to light, sun, etc., at once brings on a peculiar form of degeneration of tissue." Brooke (13) says the symmetry of the lesions is not sufficient ground for supposing a primary central causation ; some cases are to all appearance primarily central tropho-neuroses, but in the majority the fact that it spreads from one limb to its fellow, or from hands to feet, or *vice versa*, supports the theory of a reflex origin. The fact that an apparently identical lesion may be produced by the taking of arsenic is noteworthy ; but even then the congenital predisposition may be present, and the arsenic may act as the exciting cause.

* Bronson (E. B.), *Journ. of Cutan. and Ven. Dis.*, ii. p. 206, 1884.

TREATMENT.

"In congenital cases," says Crocker,* "a cure can, *à priori*, scarcely be expected." Nevertheless, various methods have been tried, and have in some instances been crowned with success. Bulkley (3) employed diachylon ointment, and Hutchinson (14) advised arsenic, tar ointments, and the use of cork soles. Unna (7) cured five members of one family by perseveringly painting the skin with a 10 to 20 per cent. solution of salicylic acid in ether. Crocker (*loc. cit.*) recommends the use of ichthyol and salicylic acid preparations.

AKROKERATOMA HEREDITARIUM.

Under the above name Max Neuburger (*Monatshefte für praktische Dermatologie*, xiii. p. 1, 1891) described a morbid condition of the skin which deserves notice at this place, for it showed certain affinities both with foetal ichthyosis and with tylosis palmæ et plantæ. The patient was a man sixty-six years of age, who had, from his early infancy, suffered from hardness of the skin of both hands and feet. His father and his eldest brother had always been similarly affected. The whole skin was rough and dry, but the extensor surfaces of the limbs were the parts most affected, especially the backs of the hands and feet, where the skin was like shagreen. All over the body the integument had a yellowish-brown colour, and the conjunctiva and buccal mucous membrane were also of a yellow hue. On the back were some seborrhœic warts, and there was a good deal of itching. Sensibility to pin-pricks was very much deadened on the feet and hands, but elsewhere it was normal. The passage from the affected skin on the back of the hands and feet to the normal on the palms and soles was quite gradual, and in this particular the disease differed markedly from tylosis palmæ et plantæ. The urine contained some albumen, but no bile pigment. Microscopic examination of the diseased skin showed a great thickening of the whole epidermis: it was from six to eight times thicker than normal. The horny layer was specially affected and had a yellowish-brown colour (in unstained

* Crocker (H. R.), *Diseases of the Skin*, 2nd edit. p. 366.

sections). In the cutis there was thinning of the connective tissue, and the elastic tissue was also diminished in quantity. The sebaceous and sudoriparous glands were much shrunken, and therefore the cutaneous secretions were almost absent. The panniculus adiposus was nearly wanting, only small clumps of fat being found in the deeper part of the subcutaneous tissue.

From a consideration of the above-mentioned microscopic appearances Neuburger concludes that the condition is not ichthyosis; he thinks it is more closely allied to the keratoma palmare et plantare hereditarium of Unna; but since in the case under review the lesion affected the extensor and not the flexor aspects of the extremities, he prefers to call it akrokeratoma hereditarium. It is true that the disease differs in many ways from ordinary ichthyosis, but it is not so unlike the cases of grave foetal ichthyosis which have been described in a previous chapter. Neuburger's patient was treated by inunctions of carbolic oil, and with a daily bath. Later, spirit of menthol (5 per cent.) was rubbed in twice daily.

TYLOMA.

The term *tyloma* has been applied to a localised variety of hyperkeratosis which results in the production of a patch of thickened and hard epidermis usually on a part of the body liable to the effect of intermittent pressure. This condition has also been described under the names "callositas," "callosity," "tylosis," "keratoma," and "durillon" (Fr.). It consists essentially in a hyperplasia of the stratum corneum of the epidermis, and has been placed, along with *tylosis palmæ et plantæ*, among the acanthoses: it is, according to Bronson (*loc. cit.*), a hyperacanthosis associated with marked hyperkeratosis. Most of the authors who have written on the subject—e.g., Dechambre (A.), Article "Durillon," in *Dict. encyclop. des Sciences méd.*, 1st series, xxx. p. 714—make no allusion to congenital cases. Doubtless acquired callosities (e.g., the well-known bunion) are so common and so striking that attention has been specially attracted to them; but the congenital and hereditary character of some cases must not be overlooked. Bland Sutton (*Evolution and Disease*, p. 19, 1890) says, "Callosities are inherited, as is shown

by the fact that the skin on the sole of a peasant's infant is thicker than that on the foot of the parson's offspring at the moment of birth;" but I do not know upon what evidence this statement, which is strongly opposed to the doctrine of Weismann, is founded. Crocker, also, in his *Diseases of the Skin* (2nd edit., p. 362, 1893), states that callosities may be congenital or acquired, and mentions the case of a mulatto woman who had flat callosities over all the first interphalangeal joints, which may have been congenital in origin.

CLAVUS (α CORN).

A corn ("cor," Fr.; "Hühnerauge," Germ.), like a callosity, consists essentially in a hyperplasia of the stratum corneum of the epidermis; but it differs from a callosity in being accompanied by a downward ingrowth which leads to atrophy of the subjacent papillæ. It is commonly acquired as the result of intermittent pressure; but, as Bland Sutton says (*Evolution and Disease*, p. 19, 1890), "We may not unreasonably attribute the readiness with which a badly fitting boot will produce corns to a tendency we inherit from our parents and grandparents." A possible congenital case was that reported by Davies-Colley (17) under the name of "Disseminated Clavus of the hands and feet;" but the fact that the author was unable to obtain the history of the patient prevents us from making any definite statement on the subject.

CORNUS CUTANEUM.

So far as I have been able to discover, no case of a congenital cutaneous horn has yet been put on record, and it is not difficult to understand why this is so. Intra-uterine life is of too short a duration to permit the formation of a horn, and the environmental conditions of the foetus are not such as to favour a growth of this kind. Since, however, warts are often congenital, and since horns are in many cases simply overgrown warts, it may safely be said that occasionally cutaneous horns are potentially present at birth. They have in rare instances developed during infancy.

The above remarks apply to the genuine cornu cutaneum, for

there are certain conditions which may be met with at birth, and to which the name "false horns" may be applied. In some of the cases of foetal ichthyosis, for example, spiny or horn-like growths have been noted; but these were not true horns. Again, preauricular appendages have sometimes a superficial resemblance to horns, but in their nature they are essentially different, being in all probability vestigial structures. I have recently seen such a case in which, at the time of birth, there was a cylindrical outgrowth in front of the tragus of the left ear, which at first sight simulated a horn. The so-called "horned men of Africa" are individuals in whom there is an exostosis or bony overgrowth of the naso-maxillary region. These exostoses are usually bilateral, and are probably always congenital. In the child seen by Strachan (*Brit. Med. Journ.*, p. 189, Jan. 27, 1894), for instance, the osseous masses were congenital, and had grown with the patient's growth. They had nothing in common with true cutaneous horns, but were simply bony projections springing from the nasal process of the superior maxilla and nasal bones.

This concludes the consideration of the idiopathic foetal skin diseases which have hyperkeratosis as their most prominent character; but it may be noted in passing, that Bronson (*Journ. of Cutaneous and Venereal Diseases*, ii. p. 206, 1884) has separated the maladies dealt with in this chapter from the hyperkeratoses proper, and has placed them in the group of the "Acanthoses," calling them "hyperacanthoses associated with marked hyperkeratosis."

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- [17. DAVIES-COLLEY (N.) — "Disseminated Clavus of the Hands and Feet," *Trans. Path. Soc. of London*, xxx. p. 451. 1879. Illustrated. In the absence of a history of the case it cannot be said whether or not the disease described by Davies-Colley was congenital; but it strikingly resembled those which have been noted above.]

CHAPTER XI.

DISEASES OF THE SKIN—Continued.

FŒTAL KERATOLYSIS : DEFINITION ; HISTORICAL NOTES ; SYNONYMS ; CLINICAL HISTORY ; DESCRIPTION OF THE FŒTUS AND FŒTAL ANNEXA ; PATHOGENESIS ; DIAGNOSIS ; PROGNOSIS ; TREATMENT ; LITERATURE.

HAVING in the previous chapter brought to a conclusion the discussion of the important diseases which have hyperkeratosis as their distinctive mark, I may now proceed to consider some cutaneous maladies which may be grouped together under the name "Keratolyses." Auspitz (*System der Hautkrankheiten*, p. 134, 1881) divided the large class of the *Epidermidoses* into several families, and in the third of these he placed " pityriasis alba " and " rubra," and " dermatitis exfoliativa infantum." To these skin diseases he gave the general name of "Keratolyses." The definition of the word "keratolysis" given in the *Illustrated Encyclopaedic Medical Dictionary* (vol. iii. p. 2068) is "a loosening or separation of the cuticle of the skin," and this very fairly describes the cutaneous lesion met with in the diseases which now fall to be considered. The first of these, to which I have given the designation "fœtal keratolysis," has been occasionally observed, and has been called by various names, e.g., "maceration in the living fœtus," etc.; the second is the malady known as "Dermatitis exfoliativa neonatorum," or "Ritter's Disease." Both these conditions are probably related in some way to the desquamation of the cuticle which occurs as a physiological phenomenon in the fœtus and new-born infant.

EPIDERMIC DESQUAMATION IN THE LIVING FŒTUS
(FŒTAL KERATOLYSIS).

As is well known, desquamation of the cuticle seen in a fœtus at birth has always been regarded as a certain sign of intra-uterine

death. A reference to the leading text-books on Midwifery and Legal Medicine will abundantly prove that this view has been universally held. It is significant, however, to find in Meymott Tidy's *Legal Medicine* (Part II. p. 285, 1883) a slightly modified statement. After referring to the general belief of writers, that peeling of the epidermis is a conclusive proof of the death of a foetus, the author goes on to say : " Still it must be noted that in *Case 52*" (that reported by Edis) "the peeling of the epidermis, occurring in a living child, was very similar, both in its nature and extent, to that which commonly happens after death." An examination of medical literature shows that the case seen by Edis was not a solitary example of ante-partum epidermic desquamation in the living foetus, and it is with it and with similar cases reported by other writers that this chapter is concerned. In all probability future investigations will show that all such cases are by no means due to one and the same cause ; there are, indeed, indications that the instances already recorded differed greatly in their pathogenesis. Doubtless the desquamation is due to a foetal disease ; but it is extremely likely that several intra-uterine maladies may give rise to this cutaneous lesion. In the meantime it is convenient to consider all the cases together under the general descriptive designation "Epidermic Desquamation in the Living Fœtus," or "Fœtal Keratolysis."

Definition.—Fœtal keratolysis may be provisionally defined as a state of abnormal looseness of attachment or of actual desquamation of the epidermis of the living foetus. It may in time be found that this morbid condition is always the result of foetal measles, scarlatina, erysipelas, syphilis, or pemphigus ; but in the cases which are grouped together in this chapter there is no *conclusive* evidence to show that any of the above-named intra-uterine maladies were present. In the meantime, therefore, I have placed this kind of foetal keratolysis among the idiopathic congenital diseases of the skin.

Historical Notes.—Saviard (1), writing in 1702, stated that he had seen several infants in which the epidermis was raised from

the underlying skin all over the body, who nevertheless survived ; and Albinus (*Academicarum annotationum*, Lib. i. p. 19, Leidæ, 1734) made reference to Saviard's observations, which were, in all probability, instances of foetal keratolysis. Schurig (2), in 1732, gathered together from earlier authors certain cases, to which he gave the name “*excoriatio fœtus*.” One of these was the observation by C. L. Göckel (“*De excoriatione fœtus in utero*,” *Miscellanea Curiosa*, etc., Dec. II., Ann. VI., Obs. cli. p. 313, 1688), in which a woman—two months before labour at the full term—suffered from a tertian fever, and gave birth to a female infant with the cuticle pustulous and excoriating, which, although in considerable danger, nevertheless, under treatment with oils, survived. From the appearance of the epidermis the child was called the “scalded one”—*dieses kind ist gebrühet auf die Welt kommen*—and the cause given was the high temperature of the liquor amnii, by which the fœtus was surrounded *in utero*. It is rather difficult to say what was the exact character of the cutaneous lesion in Göckel's case, and the same remark applies to some other observations (by Francus, Garmann, and Bartholin) also cited by Schurig. Billard (*Traité des Maladies des Enfans nouveau-nés*, 3rd ed., p. 25, Brussels, 1835) was evidently unacquainted with any instances of ante-partum desquamation in the living fœtus. Grætzer (3) considered certain of the cases quoted by Schurig to be foetal pemphigus ; whilst under the heading “*Mangel der Haut*” he placed the observations of T. Bartholin (*Hist. anat. rar.*, Cent. iii., Hist. 5, p. 15) and of Cordon (No. 2A), which were in all probability examples of foetal keratolysis, although it must be admitted that Bartholin affirmed that the whole skin was absent and the muscles exposed to view. In Cordon's case (2A) the mother thrice gave birth to infants in whom the epidermis was absent on the hands and on the legs below the knee. The affected infants were the third, fifth, and sixth ; and the first, second, and fourth had a normal skin. In his work on *Obstetric Medicine*, Blundell (4) makes the following interesting statement :—“Though the desquamation of the cuticle, however, is a strong *presumptive* argument in affirmation of the death of the fœtus, it certainly is not *demonstrative*, for cases have been related—and among the rest, one by Dr

Orme—in which the cuticle has separated in consequence of cutaneous affections, the child being alive notwithstanding. So rare, however, are these cases, that I should feel disposed to look on them as of no account, were it not that human life is at stake."

In 1840 Chowne (5) reported to the Medical Society of London a case of exfoliation of the cuticle in a new-born child, and, like Blundell, commented upon its medico-legal importance in connexion with the subject of live-birth; in the discussion which followed, Hooper said that he had seen similar cases, but they were syphilitic, and Dendy stated his belief that it was either the result of deficient nutritive power in the vessels of the skin or of foetal erysipelas. A year later Seeligmann (*De epidermidis, imprimis neonatorum, desquamatione*, Diss., Berlin) described a case of what he called "epidermic desquamation in the new-born infant;" but this case I have regarded as an instance of the mild type of foetal ichthyosis (*v. page 130*). Among the nineteen examples,* however, which Hueter (6) reported under the name "congenital desquamation" were several which ought, I think, to be regarded as "foetal keratolysis" as above defined. Bille (7) also noted a case of desquamation at birth, but there was a syphilitic history; and Sidey (8) showed to the Edinburgh Obstetrical Society yet another instance, but without details.

Edis (9), Manby (10), Charrier (11), and Hanks (12) all added to our knowledge of this rare condition; but by far the most important communication on the subject has come from the pen of Ribemont-Dessaaignes (14), who connects it closely with general dropsy of the foetus, and gives complete details of five cases. The infant, also, who was seen by Schuhl (15), showed this form of epidermic desquamation; and, finally, I (16) have observed it in some cases of general foetal dropsy which have come under my notice since the publication of the first volume of this work. In all, some thirty-five genuine examples have been put on record.

Synonyms.—Whilst Schurig (2) called the condition "excori-

* Hueter met with 17 cases (14 in male, 3 in female infants) out of 1153 children, born between 1852 and 1857, in the Marburg Maternity.

atio fœtus," and Grætzer (3) referred to it as "absence of the skin," the other authors, who described cases, generally used phrases indicating that it was a desquamation of the cuticle, and that it occurred in the living fœtus. Thus we find such expressions as "desquamation of the cuticle" (4), "exfoliation of the cuticle in the new-born child" (5), "congenital desquamation" (6), "exfoliation der Epidermis der Neugebornen" (7), "peeling off of the cuticle in the living fœtus" (8), "peeling of the epidermis in the living fœtus" (9), "epidermic desquamation of the fœtus" (10), "desquamation épidermique chez un fœtus vivant" (11), and "antepartum desquamation of the cuticle of the living fœtus" (12). Two recent writers (14 and 15), however, have given to it a name which is somewhat unfortunate—"macération chez le fœtus vivant"—for it is doubtful whether it is in any way related to the condition known as "maceration." Finally, I have, chiefly for the sake of brevity, called it "fœtal keratolysis."

CLINICAL HISTORY AND SYMPTOMATOLOGY.

A. *Maternal History*.—The *age* of the mother when she gave birth to an infant with fœtal keratolysis varied very considerably. It was 19 in Nos. 6*l* and *s*, and 12; 20 in No. 16*c*; 21 in Nos. 14*a* and *d*; 22 in No. 6*p*; 23 in No. 15; 24 in No. 6*i*; 26 in Nos. 6*m* and *q*; 27 in No. 6*h*; 28 in Nos. 6*k* and 16*a* and *b*; 29 in No. 6*c*; 30 in Nos. 6*c* and 10; 31 in Nos. 6*b*, *d*, and *o*; 32 in Nos. 6*a* and 14*e*; 35 in Nos. 6*f* and *g*, and 9; 36 in Nos. 14*b* and 16*d*; and 40 in 14*c*.

With regard to *parity*, the mother was a i.-para in nine instances (Nos. 6*f*, *g*, *l*, *m*, and *s*; 10, 12, 14*d*, and 16*c*); a ii.-para in fourteen (Nos. 6*a*, *b*, *d*, *h*, *i*, *k*, *n*, *o*, *p*, *q*, and *r*; 9, 11, and 14*a*); a iii.-para in four (Nos. 5 and 6*c*, *e*, *t*); a iv.-para in one (No. 15); a vii.-para in three (Nos. 14*b* and *e*, and 16*a*); an viii.-para in one (No. 16*b*); a ix.-para in one (No. 14*c*); and a x.-para in one also (16*d*). In No. 2A (Cordon's case) the third, fifth, and sixth pregnancies of the same mother all ended in the birth of infants with fœtal keratolysis.

The maternal *general health* was usually stated to be quite good (*e.g.*, Nos. 12, 14*c* and *d*, 15, etc.), and was specially

described as "robust" in several cases (Nos. 6c, d, e, u, etc.). In Nos. 6t and 7, however, the mother was syphilitic; and in No. 14e she had repeatedly suffered from erysipelas. In No. 6a the mother had had varioloid between the birth of her first child and the conception of the one that showed localised areas of epidermic desquamation.

The history of *previous pregnancies* did not, as a rule, throw any light upon this foetal morbid state. In Nos. 6a and d, 9, and 14a there had been one previous confinement with a normal infant; but in No. 6b the previous gestation had ended in a birth of a dead foetus. In No. 5 the two earlier pregnancies had been normal; and in Nos. 14b and e and 16a the six preceding confinements had all ended in the delivery of normal children. In No. 2A, however, the first, second, and fourth infants were normal at birth, whilst the third, fifth, and sixth suffered from congenital keratolysis; in No. 14c (a ix.-para) the first six children were healthy and the seventh and eighth were oedematous and ascitic. The earlier obstetric history in No. 15 was interesting: the mother in her first confinement gave birth to a healthy infant; her husband thereafter died from heart disease, and she married again, her second husband being a syphilitic; although she herself showed no signs of syphilis, yet she aborted at the second month; her third pregnancy ended nearly at the full term in the birth of a dead and macerated foetus showing epidermic desquamation in large lamellæ, and all the signs indicative of the advanced changes of intra-uterine death; and her fourth infant made several inspirations after birth, and then died, showing localised areas of epidermic exfoliation. The desquamation in this last infant was quite different from that in the dead and macerated child born previously.

With regard to the *characters of the pregnancy which ended in the birth of a keratolytic foetus* some information is forthcoming. In a few instances it was stated to have been quite normal (Nos. 6a, 11, and 14b); but labour occurred prematurely in Nos. 6g and 16b, at the seventh month in Nos. 6l, 14c and d, and 16a, and at the eighth month in Nos. 12, 14a and e, 15, and 16c. In two cases, however, labour was delayed; for a fortnight in No. 9, and for nearly three months (?) in No. 10. In No. 12

the mother got a severe jar eight days before labour, and thereafter lost a good deal of liquor amnii, which continued to escape till the confinement began. In No. 6*m* she suffered from "miliary fever" in pregnancy, and in No. 6*t* she had syphilis and intermittent fever during the gestation. There was hydramnios in Nos. 14*a*, *b*, *d*, and *e*; 15, and 16*d*. The mother was dropsical during pregnancy in Nos. 14*e* and 16*a* and *d*; and in No. 14*c* she suffered from dyspepsia, albuminuria, and oedema of the legs. In many instances, however, no details of the pregnancy were given. In No. 2*a* the curious statement was made that the mother had during gestation drunk specially acid things, such as vinegar; and in No. 2*b* (Göckel's case) she had suffered from malaria (tertian type).

In the reported cases of foetal keratolysis there are not always full details of the mother's *confinement*. This is unfortunate, for, of course, it leaves it an open question whether the desquamation was not in some instances due to instrumental or manual interference. Some information, however, is forthcoming. The labour was sometimes described as normal, e.g., in Nos. 5, 6*a*, 10, 14*b*, and 16*a*; and it was quick and easy in Nos. 6*d* and 11. The cord was coiled once round the neck in Nos. 6*b*, *e*, and *f*, and in No. 11. The pelvis presented in Nos. 6*c*, 12, and 16*b*; whilst it was a transverse presentation in No. 14*d*, requiring version. In certain cases the labour was delayed or difficult, and needed manual traction or the use of forceps (Nos. 6*m*, *o*, and *p*; 14*a*, *c*, and *e*; and 15). In one or two cases in which there was general foetal dropsy the infant's abdomen had to be punctured before delivery was effected. It is, therefore, evident that in some of the cases considerable force must have been employed to extract the foetus; but a scrutiny of the details of these cases shows that in many of them the desquamation of the cuticle was in regions of the body which had not been specially subjected to pressure. I believe that, whilst in a few instances (e.g., those with general dropsy) the abrasion of the cuticle was due to traction, in all of them there was at least an abnormal looseness of attachment of the epidermis as a primary cause of the separation.

B. *Paternal and Family History*.—Under this head I need

only refer again to the family prevalence of fœtal keratolysis in Cordon's cases (No. 2A). In the absence of full details one is unable, however, to exclude syphilis in this record.

C. *Infantile History*.—In all the recorded cases the fœtus was alive at birth, or at least very shortly before its expulsion from the maternal passages, and in certain instances the infant survived its birth by some minutes, hours, or even days.

With regard to *sex*, the fœtus was a male in 19 cases (Nos. 6a, b, e, f, g, k, l, m, o, p, q, r, s, and t; 9, 10, 14a and d, and 16b); and a female in 9 cases (Nos. 6c, d, h, and n; 14b, c, and e; and 16a and c). The predominance of male infants may point to the large size of the fœtus as a causal factor in the production of keratolysis.

The affected infant was by no means always large in size: thus, whilst it weighed 9 lbs. in No. 6r, 8 lbs. in No. 9, 3830 grammes in No. 14c, and 3300 grammes in No. 14e, its weight was 3½ lbs. in Nos. 6b and g, 4½ lbs. in Nos. 6f, i, and l, 4¾ lbs. in No. 6d, 6 lbs. in No. 6a, 6½ lbs. in No. 6k, 6½ lbs. in No. 6t, and 7 lbs. in No. 6e. In length it varied from 16 to 20 inches.

The infant was often weakly,—especially, of course, in the cases in which labour took place prematurely; but sometimes it was robust. It died during labour in Nos. 6l, 14a and b; in a few minutes after birth in Nos. 8, 14d and e, 15, and 16c; in half an hour in No. 12; in one hour in Nos. 5 and 6f; in five and a half hours in No. 6g; and in "some hours" in Nos. 9 and 6b. It was alive when the mother left the Maternity Hospital in most of Hueter's cases, viz., in Nos. 6a, c, d, e, k, m, n, o, p, q, r, s, and t; and it also survived in Nos. 1, 2a, 10, and 11.

DESCRIPTION OF THE FœTUS.

In all the recorded cases there was epidermic desquamation, but this morbid state of cuticle varied greatly in extent. It was described as universal (affecting the whole body) in Nos. 1, 2, 3, 6m and n, 9, 10 (?), and 11; in No. 5 it was nearly as widespread, for the arms and hands, legs and feet, neck and back, and even the head were all described as in a state of desquamation; but in most of the other cases the lesion was much more localised. In Nos. 2A, 6a, f, g, h, i, l, p, and t, and 14d the limbs

alone were affected ; and in Nos. 6*i* and *t* the palms and soles were the only regions to show the morbid condition. In No. 6*s* the hands alone were affected, in No. 14*e* the arms alone, and in Nos. 12 and 16*b* the change was seen only on the legs. In No. 6*c* the desquamation was limited to the left side of the chest ; in No. 14*d* the chest, hands, and feet suffered ; in No. 14*e* the chest and abdomen showed the change ; in No. 14*b* the skin of the cheeks and of the dorsal surface of the hands and feet was in a state of keratolysis ; and in No. 14*c* the change was limited to the chest, neck, and abdomen. In No. 6*o* there was desquamation on the hands, feet, and scrotum ; on the hands, feet, scrotum, and face in No. 6*k* ; on the shoulders, chest, neck, iliac regions, and penis in No. 14*a* ; on the limbs, face, and penis in No. 15 ; and on the lower part of the abdomen alone in Nos. 16*a* and *c*.

The skin showed other changes in addition to the desquamation in a few instances. In Nos. 5 and 6*a* cicatricial spots were visible, in the latter case being limited to the head. In one or two cases (*e.g.*, No. 9) the whole integument was described as of a dusky blue colour. In Nos. 6*h* and *m* there were "miliary vesicles" to be seen, and in Nos. 6*k* and *o* the lesions of pemphigus appeared soon after birth. In a group of cases (Nos. 14*a*, *b*, *c*, *d*, and *e*, and 16*a*, *b*, *c*, and *d*) there were also epidermic blebs containing clear fluid, and a state of general anasarca of the subcutaneous tissue of the foetus.

In many instances no information regarding the vernix caseosa was forthcoming ; but in Nos. 6*a* and *e* it was absent ; in No. 6*k* it was small in amount ; in No. 6*b* it was present ; and in Nos. 6*o* and *p* it was copious.

Full details were not often given with regard to the nature of the epidermic desquamation. In Nos. 6*h* and *k* it was described as furfuraceous, whilst in No. 6*n* it—the cuticle—separated in large membranous portions. In No. 9 the epidermis came away in large flakes, being more adherent in some parts than others, but still peeling readily all over the body. Much the same condition was noted in No. 10, where the raw surface of the cutis was left exposed ; and in No. 11, in which the desquamation was universal, the epidermis of the foot came off like a glove. In

Nos. 9, 11, and 12 the appearances were described as quite similar to those in a macerated foetus. In a group of cases (Nos. 14*a*, *b*, *c*, *d*, and *e*, and 16*a*, *b*, *c*, and *d*) in which the desquamation was accompanied by general foetal dropsy, the epidermis was raised at places by a clear—not sanguinolent—serum, whilst in other areas it was absent, and the underlying dermis, which had a pale rose or salmon tint, was exposed. From the character of the serum in the vesicles, and from the colour of the exposed dermis, it was hoped that it might be possible to distinguish desquamation in the living foetus from the macerative changes seen in the dead infant; but in No. 15 Schuhl noted areas in which the dermis was pale rose in colour, and others in which it had a bright red tint, so that both varieties of desquamation were there present.

In the cases in which the infant died shortly after (or during) birth an autopsy was rarely made. In No. 6*b*, however, it was stated that no pathological changes were met with save pulmonary atelectasis, spinal and cerebral anaemia, and a haemorrhage below the tentorium cerebelli. In Ribemont-Dessaaignes' specimens (Nos. 14*a*, *b*, *c*, *d*, and *e*) and in my own (No. 16) a complete post-mortem examination was made; but the morbid conditions, which were numerous and varied, were doubtless associated with the state of general anasarca and dropsy of the body-cavities rather than with the epidermic desquamation.

DESCRIPTION OF THE FœTAL ANNEXA.

In only a few instances were any details given concerning the foetal annexa. The umbilical cord was twisted round the neck of the foetus in Nos. 6*b*, *e*, and 1; it had a green and red colour, and was flattened like a ribbon in No. 11; and it was short in No. 14*a*. The foetal membranes had a macerated appearance (as had the cord) in No. 11. There was hydramnios in Nos. 12, 14*a*, *b*, *d*, and *e*; 15, and 16; but the liquor amnii was described as normal in No. 11. In the cases in which there was general foetal dropsy (Nos. 14 and 16) the placenta was large, oedematous, and friable; and in No. 15 it was large, but otherwise normal. In No. 10 it was small, ragged, and quite "rotten." In one or two instances it was described as showing fibrous areas.

Unfortunately the absence of details in regard to the foetal annexa, in the great majority of cases, prevents us from arriving at any conclusion with regard to the bearing of placental disease, etc., upon the desquamation of the infant.

NATURE OF THE MORBID PROCESS.

In all the observations that have been referred to above there seems to be no doubt that epidermic desquamation existed in the infant at the moment of birth,—was, in a word, truly congenital; but in only a few instances was there any indication regarding the nature and mode of origin of the keratolysis. Many of the observers did not, indeed, discuss this question at all. It may, however, be well to pass in review the opinions of those who did.

Göckel (*2b*) regarded it as certain that the condition of the cuticle in the infant seen by him was due to the action of the liquor amnii in which it floated *in utero*; since the mother suffered from fever, the temperature of this fluid would also be raised, and thus the foetus would be scalded. Blundell (*4*), writing long after Göckel, evidently considered the desquamation as caused by cutaneous affections in the foetus, but does not specify any. In the discussion that followed the reading of Chowne's paper (*5*), Hooper stated that he had seen similar cases, but in syphilitic infants, and thought that in the case reported the low state of vitality, and probably some unusual state of the liquor amnii, might in some measure serve to explain the exfoliation. Dendy, on the same occasion, affirmed that it was either the result of deficient nutritive power in the vessels of the epidermis, or of foetal erysipelas. Hueter (*6*) seems to have regarded congenital desquamation as due to many conditions (syphilis, foetal measles, scarlet fever, etc., protracted gestation, pemphigus, etc.); whilst Charrier (*11*) discussed the question whether the foetus could become macerated in the amniotic fluid before death, but thought that it was more rational to connect the epidermic desquamation with some morbid state of the foetus occurring late in intra-uterine life. Hanks (*12*) was of opinion that the exfoliation, which in his patient affected only the lower limbs, was due to obstruction of the foetal circulation

from the marked flexion of the thighs which existed ; and in the discussion which followed his communication, Jacobi stated that it might be due to the compression of the femoral arteries thus brought about, or to a pemphigoid dermatitis, probably syphilitic in nature ; and Mundé also suggested syphilitic pemphigus as the cause. Ribemont-Dessaignes (14) is, I think, correct in his statement that in foetuses with general anasarca the appearances of maceration are caused by the rupture of little epidermic vesicles containing opalescent fluid. With regard to the cases of non-dropsical infants born alive, he was forced to admit that it was more difficult to find a cause for the exfoliation, but thought that it might arise in some circulatory disturbance leading to slight infiltration of the cutaneous tissues without actual oedema.

The diversity of opinion which has been thus shown to exist regarding the nature of foetal keratolysis fully warrants my assertion, that it is in all probability a symptom of various morbid states rather than a distinct pathological entity. I do not think that congenital desquamation of the cuticle of the living infant is ever truly physiological ; it is, in my opinion, to be regarded as the result of a pathological process. It is quite true that exfoliation of the cuticle and depilation go on during intra-uterine life, for it is impossible otherwise to explain the composition of the vernix caseosa and the contents of the intestine, and it is also true that desquamation normally commences from two to three or even five to six days after the birth of the neonatus ; but the keratolytic state of the epidermis seen at the time of birth is a morbid condition.

In one group of cases it undoubtedly has to do with foetal syphilis, and something will be said of this form of it when that disease is considered. In another set of instances it is the consequence of intra-uterine measles or scarlet fever, a fact which I have, I think, conclusively shown in two recent papers.* In the cases, however, which have been referred to in this chapter, foetal syphilis, measles, and scarlatina were practically excluded,

* "Congenital Measles," *Archives of Pediatrics*, April 1893, and "A Case of Scarlet Fever in Pregnancy with Infection of the Fœtus," *Edin. Med. Journ.*, July 1893.

and it is, therefore, necessary to look for other causes of this unusual state of the epidermis. Certain possible explanations are forthcoming.

In *one* group of cases there can be no doubt that the desquamation of the cuticle seen at birth is due to foetal anasarca. In several cases I have convinced myself that this is so. On account of the dropsical condition of the skin little collections of clear fluid occur under the epidermis and raise it in the form of vesicles. During labour the pressure of surrounding parts leads to rupture of some of these vesicles and removal of the cuticle, and thus denuded areas of skin are produced; in other less exposed regions of the body the vesicles may remain intact and be recognised as such at the time of birth. Ribemont-Dessaignes (14) states that from the character of the vesicle contents and from the colour of the denuded skin it is possible to distinguish "maceration" in the living fœtus from the genuine macerative post-mortem change; but whilst this may be true of the desquamation associated with general foetal dropsy, it is not always so in other cases, for in the infant seen by Schuhl (15) both varieties were present at the same time.

In a *second* group of cases the cause of the desquamation is to be looked for in foetal pemphigus. In several of the instances reported by Hueter (6), pemphigus was developed a few days after birth, and it is, therefore, fair to suppose that it may have existed *in utero*, and that the denuded areas were due to the rupture of the characteristic blebs during labour. The pemphigus need not necessarily have been syphilitic in nature.

It is possible that in a *third* group should be placed certain cases in which, on account of the protraction of gestation beyond its usual term, the fœtus comes over-ripe into the world, and shows a condition of the integument normal as to the date of its occurrence, but abnormal in the fact that it has taken place *in utero*.

In yet another (*fourth*) series of instances it may be that from some disturbance of the nutrition of the skin the cuticle loses its close attachment with the underlying cutis, and so is thrown off in the form of flakes and scales. The cause of the nutritional

disturbance we can only guess at : it may be due to compression of a large bloodvessel,—*e.g.*, the femoral, as in No. 12,—or to foetal erysipelas, or to some abnormal condition of the circumambient liquor amnii.

Finally, it may be necessary to relegate some of the cases to a *fifth* category and to consider them as instances of the development *in utero* of the mildest type of foetal ichthyosis. Before, however, such a conclusion could be arrived at, it would be necessary that the condition should be to some extent permanent.

Possibly there may yet be other causes which lead to the occurrence of desquamation of the cuticle in the living foetus ; but until the mechanism of the normal post-partum neonatal keratolysis is fully understood, the pathogenesis of the foetal variety must in many points remain obscure.

DIAGNOSIS, PROGNOSIS, AND TREATMENT.

Maceration in the dead foetus may possibly be confused with foetal keratolysis as above defined, especially if the observer be not present when the infant is born, and if, also, death occur soon after its expulsion from the maternal passages. In such a case one must rely upon the absence of the other signs of post-mortem maceration, and upon the presence of the changes in the infant's lungs, heart, etc., which point to the temporary establishment of an independent existence. Ribemont-Dessaignes' test is not always a certain one ; for whilst as a general rule the denuded areas are bright red in post-mortem maceration, and of a pale salmon tint in foetal keratolysis, this difference does not constantly exist. When vesicles are present it is generally true that their contents are sanguinolent in the true macerative change, and clear or opalescent in the keratolytic ; but this also, is not invariable. The distinction between the different varieties of foetal keratolysis must in the meantime be founded upon the history of the case, the presence or absence of syphilis in the parents, and upon the concomitant skin affections,—*e.g.*, pemphigus, which may be noted either at birth or in the week following.

The prognosis is not necessarily always grave, for in many of

the cases recorded by Hueter (6) the infants left the hospital in a perfectly healthy state ; at the same time it must always be very guarded.

The treatment consists in the protection (by ointments, vaseline, cotton wool, and the like) of the denuded areas of skin from the effects of cold, from irritation, and from septic infection ; but, as is usual with foetal morbid states, no very great therapeutic successes have to be recorded in this direction.

The medico-legal importance of the fact that an infant may be born alive and nevertheless show the condition of the cuticle that has almost constantly been associated with the maceration that follows intra-uterine death has been already referred to and must not be forgotten, for, as has been said by Blundell (4), human life may be at stake (*e.g.*, in a trial for infanticide).

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CHAPTER XII.

DISEASES OF THE SKIN—Continued.

DERMATITIS EXFOLIATIVA OR KERATOLYSIS NEONATORUM: DEFINITION; HISTORICAL NOTES
SYNONYMS; SYMPTOMATOLOGY; PATHOLOGY AND PATHOGENESIS; DIAGNOSIS; PROGNOSIS;
TREATMENT. PERIODICAL KERATOLYSIS: SYNONYMS; HISTORICAL NOTES; SYMPTOMATOLOGY;
NATURE AND MODE OF ORIGIN; LITERATURE.

THERE is a cutaneous affection to which the name Ritter's disease has often been given, which must, I think, be considered here, for it would seem to stand in the same sort of relationship to foetal keratolysis as neonatal sclerema does to true congenital sclerema. Like foetal keratolysis, it is evidently connected in a more or less definite manner with the normal exfoliation of the cuticle of the new-born infant. Possibly it may be permissible to regard foetal keratolysis as a premature and excessive form of the desquamation of the cuticle which normally occurs in the neonatal state, whilst Ritter's disease is considered to be simply an exaggeration of that process. They may both be looked upon as abnormally marked epidermic exfoliations, with this difference, that the one is evident at birth, whilst the other comes on a few days later.

Definition.—It is impossible, in the present state of our knowledge, to frame a satisfactory definition of dermatitis exfoliativa neonatorum, as Ritter's disease is commonly called; but bearing in mind its most prominent clinical feature, it may be stated to be a general and excessive desquamation of the cuticle, occurring in the neonatal state; it is, in fact, a keratolysis of the new-born (keratolysis neonatorum). Elliot (17), the greatest authority upon this disease in the English-speaking medical world, ventures on a somewhat fuller definition. According to this dermatologist, it is "an acute disease of the new-born, characterised by progressive implication of the cutaneous surface

and excessive epidermic exfoliation, and accompanied at times by the formation of vesicles and bullæ." He also adds: "Though the general functional health is unaffected, the rate of mortality from secondary or other causes is nearly fifty per cent."

Historical Notes.—It was not till the year 1878, when G. Ritter von Rittershain (1) published his classical monograph entitled *Die exfoliative Dermatitis jüngerer Säuglinge*, that keratolysis neonatorum came to be recognised as a distinct disease. Kaposi (4), in 1881, claimed that the malady had been already described many years previously by Billard, von Baer, Hueter, and Bille; but certainly none of these writers specially emphasized its existence as a separate disease, and further, it seems to me that they were acquainted with the foetal rather than with the neonatal type (*vide Fœtal Keratolysis*, p. 191 *et seq.*). Doubtless cases of it occurred prior to Ritter's time, but they were most probably classified by their observers as anomalous varieties of pemphigus neonatorum.

The history of the way in which Ritter gradually was led to formulate his views on the disease which bears his name is interesting. During the ten years from 1868 to 1878 he had frequent opportunities of watching the malady, for it was then epidemic in the Foundling Asylum at Prague, and he saw, in all, 297 cases of it. His first opinions regarding it were stated in 1868 (*Jahresbericht der kon. böhm. Landes-Findelanstalt*), and two years later he again discussed the condition under the name of "Dermatitis erysipelatosa" in the *Oesterr. Jahrbuch für Pädiatrik*, 1 Jahrgg. 1870, Bd. i. p. 26. It was not, however, till 1878 that he fully described the malady, and as the result of his large experience gave to it the designation "Dermatitis exfoliativa neonatorum," or "Die exfoliative Dermatitis jüngerer Säuglinge."

Whilst Ritter was engaged in watching the progress of his cases in Prague, seven instances were put on record in one of the political newspapers of that town (*Bohemia*, No. 286, 1868). The quotation was as follows: "In einem Dorfe des Stettiner Kreises ist in diesem Herbste eine eigenthümliche Krankheit, die sogenannte Blasenfaulniss, aufgetreten, welche sich darin

äusserte, dass bei allen Kindern, welche von Anfang August bis 20 September geboren wurden, in wenig Tagen eine vollständige Enthäutung und von 7 Kindern 5 starben." These cases were known also to G. Behrend (3), who was the first to write on the disease after the appearance of Ritter's article. Neither he nor Ritter saw the cases; but to the former an account of them was communicated by an eye-witness, Dr Litten of Neu-Stettin in Pomerania. Dr Litten regarded them as examples of acute pemphigus foliaceus, and from his description Behrend concluded that they were the same as Ritter's disease, and that, therefore, the latter was simply acute pemphigus foliaceus (Cazenave) affecting the new-born infant.

A true case of dermatitis exfoliativa neonatorum seems to have been that described in 1878 by C. Boeck (2) under the name of pemphigus neonatorum; but notwithstanding the appearance of the monograph of Ritter and its criticism by Behrend, extremely few instances of the disease have since been put on record. Kaposi (4) and Auspitz (5) both discussed the subject in 1881: the former agreed with Ritter in separating the malady from pemphigus foliaceus, but differed from him in regarding it as a simple exaggeration of the normal desquamation of the new-born and not as a pyæmic process; whilst the latter placed it with pityriasis alba and rubra in the group of the keratolyses, and considered it to be due to an atrophic disease of the rete Malpighii.

In 1883, Bohn (6) described Ritter's malady at some length under the name "Pemphigoide Zustände der Neugeborenen." He had seen a few cases in private practice, and believed it to be a dermatitis. In the same year Brocq (7) and Weyl (8) discussed its manifestations, the former regarding it as a form of pemphigus; but it cannot be allowed that either these writers or Bohn threw much light upon its nature. In 1884, however, Caspary (9) made an important contribution to the subject, for he described a typical case, pointed out that it could not be regarded as inflammatory in nature, and advanced the theory that it might be due to an acute disturbance in the nutrition of the superficial non-vascular layers of the epidermis, with secondary hyperæmia of the cutis.

In 1888, G. T. Elliot of New York (10) reported fully two cases which he had seen, and discussed at considerable length the history, pathology, and clinical manifestations of the disease; he was inclined to adopt Caspary's theory of its origin, but as he had not been able to make a post-mortem examination in any case, he hesitated to advance a definite opinion. In the same year B. Whitford (11) reported "a case of dermatitis exfoliata in a new-born infant" to the Rhode Island Medical Society; but I have not been able to refer to his article in the original, and recent writers do not even mention it.

Besnier and Doyon (12) in their copiously annotated translation of Kaposi's work, do not add much to our knowledge of Ritter's disease; but they recorded a congenital instance of pemphigus foliaceus, and stated their belief that it was to be regarded as quite distinct from dermatitis exfoliativa neonatorum. Max Joseph (13) also discussed the disease in his text-book, but did not offer any novel suggestions as to its nature and mode of origin.

In 1892, Raymond and Barbe (14) reported the case of an infant which showed on the tenth day a diffuse redness of the skin with universal epidermic desquamation; but whilst the authors regarded it as Ritter's disease, Brocq, Vidal, and Besnier seem to have had grave doubts on the subject. In the following year Crocker (15) considered the malady in his text-book on *Diseases of the Skin*, placing it with pityriasis rubra and epidemic exfoliative dermatitis in the class of the "exudationes." In 1893, also, Max Runge (16), in the second edition of his work on Neonatal Diseases, referred very briefly to it in the chapter devoted to pemphigus neonatorum.

In 1894, Elliot (17) again considered Ritter's disease at some length, and gave his later opinions on its nature, stating that he had seen in all six cases of it. He referred to a case shown by Hallopeau to the Dermatological Society of Paris in 1892, but omitted to give the reference, and I am inclined to believe that he was thinking of the example met with by Raymond and Barbe, for I can find no allusion to any instance reported in that year by Hallopeau. Elliot was of opinion that the

disease had probably a parasitic mode of origin, and cited in support of this theory Richl's discovery of a fungus in it.

Synonyms.—Several names have been given to this affection, but those which have been most commonly employed are "Dermatitis exfoliativa neonatorum" and "Ritter's disease." Ritter himself first called it "Dermatitis cysipelatosa," but abandoned this term later for "Exfoliative Dermatitis der Säuglinge." Bohn, Caspary, Elliot, and others have either adopted in a slightly modified form this second designation, or have complimented the first investigator of the disease by calling it simply "Ritter's disease." According to some authors, it ought to be called a variety of pemphigus neonatorum; and I have ventured to give to it the designation "Keratolysis neonatorum."

Symptomatology.—Ritter's description of the clinical manifestations of this disease, founded as it was upon an experience which was quite unique, remains up to the present the classical one. Upon Ritter's monograph, and upon the excellently described cases met with by Elliot (10, 17), the following account of the symptomatology of the malady is based. I have not personally seen a case of keratolysis neonatorum; but my friend Dr Wm. Fordyce has given me details of an infant under his care some time ago which seems, from his description, to have suffered from Ritter's disease.

It does not appear that there is anything in the history of the parents, or in the circumstances of the pregnancy and confinement, that predisposes the infant to the development of dermatitis exfoliativa. It is noteworthy, however, that in Elliot's second case the mother, a Jewess, stated that during the last four or five months of her pregnancy she had suffered from an exceedingly itchy eruption of little blisters upon the extensor surface of the arms and thighs, which had spontaneously disappeared two weeks before labour. There is usually an entire absence of any indication of a syphilitic family history. The infants at birth are perfectly healthy in appearance, and are generally full time, but they may occasionally be premature.

According to Ritter's statistics, male infants are more frequently affected than female, in the proportion of 165 of the former (55·5 per cent.) to 132 of the latter (44·4 per cent.). Of the six cases seen by Elliot (10, 17) the sex of two only is stated, and they were both females. The infant described by Raymond and Barbe (14) was a male. Probably sex has little or no influence upon the development of the disease.

Ritter's disease usually attacks the infant at a definite age. It is rarely met with before the end of the first week of life; it is common in the second week; it may be found commencing in the third, fourth, and even in the fifth week; but is extremely uncommon after that date. It is therefore perfectly appropriate to term it a *neonatal* malady.

Its commencement is usually sudden; but Ritter describes a prodromal stage in which there is seen a dry scaly condition of the epidermis which has persisted after the so-called physiological desquamation of the new-born has taken place. He has also divided the course of the disease into four further stages, viz.—(1), a stage of erythema and exudation; (2), a stage of exfoliation and drying; (3), a stage of re-integration of the epidermis; and (4), a stage of sequelæ,—e.g., boils. This division, however, is largely artificial, and one stage often passes almost insensibly into another.

The first phenomenon noticed is the sudden development of a redness of the skin, varying in tint from a light to a dark purple-red. This usually appears first about the lower part of the face, and is generally accompanied by fissures at the angles of the mouth, congestion of the nasal and buccal mucous membranes, and injection and reddening of the conjunctivæ.

Sometimes the redness first makes its appearance over the buttocks or elsewhere, whilst in other cases it may be practically universal from the very first. When it appears in localised areas it soon spreads to surrounding parts, and so in a short time (five days in Elliot's second patient) becomes general over the whole body. The redness usually last invades the arms and legs. The infant remains well nourished, takes the breast, and there is no elevation of temperature.

The stage of exfoliation follows quickly upon that of ery-

thema ; in fact, the redness is often seen invading one area of the body whilst desquamation is going on in another. Crusts form on the lower part of the face, and under these the skin becomes deeply fissured. The epidermis over the whole body becomes thickened, and is raised up from the cutis by a thin layer of fluid exudation. The cuticle is soon thrown off in large masses; the exposed cutis is seen to have a dark red colour, and the infant looks as if he were the subject of an extensive burn. Sometimes a thin yellowish crust forms on the exposed cutis, which becomes dry and adheres firmly, especially by its edges. On the hands the epidermis sometimes separates almost like a glove, and on the feet it peels off in large flakes. Even in this stage there is little or no constitutional disturbance.

The stages of erythema and desquamation, which have been described above, varied considerably in their manifestations in different cases. Sometimes the skin remains remarkably dry throughout the whole course of the disease. Then no large masses of epidermis are thrown off, but the skin becomes fissured and has a parchment-like dryness. Elliot (10) gives a very good description of the appearances seen in his first patient. "The desquamation of the epidermis, which had begun on the face, had followed progressively the march of the redness, and the surface was covered with large thin lamellæ and scales, which came off freely on the slightest rubbing. When the baby was at rest, the body appeared as though covered with a silvery, slightly wrinkled veil, which was fissured in every direction into squamæ of all sizes and shapes, the free edges of which were rolled up and slightly loosened from the underlying surface. On the face and scalp the scales were more adherent, but on the extremities they were very marked, though the redness was not as intense as on the other portions of the body. There was no moisture found beneath any of them, but only a bright red and slightly glazed surface. Under the microscope the squamæ were found to consist entirely of horny epidermis."

Sometimes, again, thickly scattered miliary vesicles are seen, chiefly situated on the forehead and extending to the scalp. In other cases the eruption resembles an eczema ; and in still

other cases the epidermis is raised in vesicles and bullæ like a pemphigus.

All these changes usually occur within a week of the first appearance of the erythema. When there was no exudation the exfoliation did not take place so rapidly, and the absence of the exudation was, according to Ritter, met with in children whose nutrition had suffered from other causes.

The next stage (Ritter's fourth stadium) is that of re-formation or re-integration of the cuticle. Along with a fading of the erythema (usually delayed on the extremities) there is a regeneration of the cuticle, often very rapid in its course. The skin, however, may remain scaly and irritable for some time. Where there has been no exudation the process of repair is often delayed; and the infant may die from complications before the epidermic regeneration is completed.

Often the whole process is completed without severe constitutional symptoms. There is commonly an entire absence of fever, as Caspary (9) was careful to point out; the infant does not lose weight, it may even gain it; and there is no diarrhoea. In other cases, however, pneumonia, green stools, even colliquative diarrhoea, may occur as complications; or the disease may be followed by a marasmic condition, the infant becoming cachectic and marasmic. Sometimes relapses occur, usually about ten days after the first attack; but they seldom prove dangerous. When complications occur, systemic disturbance is of course at once set up.

In a final stage Ritter places the sequelæ of the disease, which are usually of the nature of boils or of abscesses, in the subcutaneous tissue. These sometimes pass on into extensive phlegmonous infiltration with gangrene and sepsis, and death follows from intercurrent pneumonia or colliquative diarrhoea. Eczema may also be noted as a sequela. In one of Elliot's cases there was a severe affection of the eyes. This he believed to be primarily only an exaggerated form of the ocular changes occurring so frequently in Ritter's disease,—generally, however, in a minor degree. There is exfoliation of the epithelial layer of the cornea, just in the same way as there is desquamation of the epidermis. In the case referred

to, the denuded cornea was exposed to irritation from the movements of the eyelids, dust, etc., and perforation of the cornea, with prolapse of the iris and haemorrhage, was the result.

Pathology and Pathogenesis.—The morbid anatomy of Ritter's disease seems to consist simply in hyperæmia of the cutis, with looseness of attachment and subsequent desquamation of the epidermis, and with the occasional presence of vesicles and bullæ; but with regard to the question of pathogenesis there exists great diversity of opinion. The various theories of its mode of origin may be briefly enumerated.

Ritter (1) was of opinion that dermatitis exfoliativa neonatorum was essentially a pyæmic manifestation, and he advanced in favour of this view the fact that the disease was frequently followed by inflammatory and purulent formations. Elliot (17), however, regards Ritter's opinion as far from satisfactory, "owing to the difficulty in imagining a pyæmic process without a purulent focus as a starting-point, or one so extensive and yet localised entirely in the superficial layers of the skin, or one running such an acute course without any elevation of temperature." The argument, based upon the inflammatory sequelæ, also is weakened by the fact that they are sequelæ and not antecedents. At the same time the gradual disappearance of the disease from the Prague Hospital, in association with an improvement of its hygienic conditions, as pointed out by Epstein (*Archiv fur Kinderheilkunde*, vii. p. 98, 1886) would seem to support Ritter's contention. Further, the frequent presence of a subnormal temperature in the neonatal state might mask the presence of fever in these cases, just as it sometimes does in pneumonia at this age.

Behrend (3) regarded the malady as pemphigus foliaceus (of Cazenave) occurring in the new-born; and although Dr Litten's cases may have belonged to this category, there is nothing to prove that they were of the same nature as those seen by Ritter. Still, the theory that the disease was pemphigus, or at any rate pemphigoid, has been a popular one, and has been held by Bohn (6), Brocq (7), Max Runge (16), and others.

Bohn (6) also thought that it was essentially a dermatitis,

a local process brought about by some external influence ; but Caspary (9) believed that the absence of fever negatived this opinion, and was further persuaded that it was not a form of pemphigus. The latter writer did not, however, throw much light upon the subject when he stated that it was "an epidermolysis of unknown nature with secondary hyperæmia of the cutis,—possibly an acute disturbance of nutrition in those external layers of the skin which do not contain bloodvessels."

Kaposi (4 and 12) considered it as an exaggeration of the physiological exfoliation of the epidermis in the new-born, a belief in which I am inclined to share ; but he also stated that Riehl had in some cases of the disease found a fungus with long and thin mycelia, which he regarded as its cause. With regard to Riehl's suggestion, Elliot (17) says that although wanting corroboration, it is the most satisfactory that has been given ; adding, however, that he had been unable to find the fungus in one case examined by himself.

Now, the nature of the normal desquamation of the new-born infant is not yet known. It may be that it is the external sign of a mild form of pyæmia ; it may also be that it is due to a fungus, or it may simply be caused by drying of the epidermis which has long been soaked in the liquor amnii ; but I am strongly of opinion that Ritter's disease is closely allied both to it and to some, at any rate, of the cases that I have called foetal keratolysis. It may also be of the same nature as the so-called scarlatinaform rashes sometimes seen in puerperal women. It would seem that just before and just after birth the attachment of the epidermis to the underlying cutis is not very firm, and that various causes may easily cause its separation and exfoliation. Even the "physiological" desquamation in the new-born deserves further investigation.

Diagnosis.—Elliot (10, 17) considers in considerable detail the various affections which may be confounded with Ritter's disease, and points out that we must trust to the age of the patient, the mode of extension of the lesion, its superficial seat, its rapid but non-febrile course, and its desquamative characters, in order to distinguish it from them.

From the *pityriasis rubra of Hebra* it can easily be separated, for this, whilst being essentially a disease of adult life, takes months and even years to become widespread. The *dermatitis exfoliativa* of Erasmus Wilson also affects the adult is accompanied by grave constitutional symptoms, and after a course of a few months commonly terminates favourably. From *erythema neonatorum*, Ritter's disease must also be distinguished, and from the *physiological desquamation of the new-born*; but it may also be that these maladies differ from it in degree rather than in kind. *Erysipelas neonatorum* is usually localised to the periumbilical region of the body, and is, furthermore, accompanied by grave systemic disturbance, and by marked local swelling of the affected area.

Generalised acute *eczema* sometimes occurs in infants; but unlike Ritter's disease it is accompanied by fever, by irritation leading to itching, by a redness which is the result of cutaneous lesions of a polymorphous kind which are primarily separate, by the characteristic crusts and weeping surfaces, and by a tendency to become chronic.

Pemphigus has some characters in common with neonatal keratolysis; but it is essentially a bullous affection, whilst Ritter's disease either shows no bullæ, or shows them only as secondary formations. *Pemphigus foliaceus* is a chronic disease of adult life; and *pemphigus simplex acutus neonatorum* appears at an earlier date than Ritter's disease, shows discrete bullæ and not a diffuse redness as its primary manifestation, and has a tendency to form successive crops.

Desquamation due to *scarlet fever* or to *measles* acquired either *in utero* or immediately after birth might at first sight strongly resemble the conditions seen in *dermatitis exfoliativa neonatorum*; but the history of the case, the high temperature, and the presence of characteristic throat or eye manifestations, ought to serve as reliable means of diagnosis.

Prognosis.—The fact that Ritter lost 50·9 per cent. of the male, and 46·2 per cent. of the female infants affected with the disease which bears his name, is sufficient proof that a very grave prognosis ought always to be given. The disease may

prove fatal by the intensity of its initial lesions, as sometimes happens in grave cases of scarlet fever; but more often the cause of death is to be found in exhaustion, marasmus, secondary diarrhoea, pneumonia, or blood-poisoning, and perhaps in the great loss of body-heat due to the exfoliation of the epidermic layer of the skin. Of Elliot's six cases, four died, and the fate of another was uncertain: one succumbed to haemorrhage from the bloodvessels of the prolapsed irides, another to secondary gastro-enteritis, a third to inanition from loss of body-heat, and the fourth to general debility during a relapse. The tendency to relapses, to complications, and to dangerous sequelæ must not be lost sight of in forming a prognosis.

Treatment.—Ritter's disease can, of course, only be treated symptomatically, and necessarily often ineffectually. The treatment must be both general and local. Local measures consist in the protection of the desquamating surface by cotton wool, and by the application of oils and fats; and on account of the possible causation of the disease by a fungus, Elliot specially recommends ichthyol, boric acid, or resorcin, for their antiseptic and parasiticidal action. The general health should be supported by tonics, regular nourishment, and preferably by the breast milk, and good hygiene. Complications and sequelæ, when they occur, will demand their appropriate treatment, boils and abscesses requiring antiseptic dressing.

PERIODICAL KERATOLYSIS.

Under the names "Keratolysis" and "Deciduous Skin," Radcliffe Crocker (*Diseases of the Skin*, 2nd edit., p. 240, 1893) mentions two recorded instances of a curious affection in which there is a periodical casting-off of the cuticle, and quite recently Sangster (22) has reported an example of a closely allied morbid state which would seem to connect it with dermatitis exfoliativa neonatorum and foetal keratolysis. It is therefore necessary that some reference be made to it in this work.

Synonyms.—In addition to the names "Deciduous Skin" and

"Keratolysis," this cutaneous affection has been called "recurrent exfoliative erythema" by Fox (18), "periodical peeling of the cuticle" by Chevallier Preston (19), "annual shedding of the epidermis" by Sligh (21), and "congenital exfoliation of the skin (keratolysis exfoliativa?)" by Sangster (22). Several of these names are definitions rather than designations; and with regard to keratolysis exfoliativa, exception must be taken on account of its tautological character. "Deciduous Skin" suggests the idea of the separation of more than the epidermis; and, on the whole, I prefer the term "Periodical Keratolysis," as expressing shortly the nature and most important character of the cutaneous lesion.

Historical Notes.—Doubtless cases of this disease have occurred in the past, and possibly may be noticed under other names in older writers; but the first instance I have found of it is that reported by Fox (18) in 1879. In 1881 Preston (19) saw an example of it in a woman in New Zealand, and ten years later Frank and Sandford (20) noted a case in a male subject in Chicago. Sligh (21) reported the same case in 1893, and Sangster (22) has recently (February 1895) put on record a "congenital" instance of an allied affection seen by him in London at the Charing Cross Hospital.

Symptomatology.—As the recorded cases are few in number the clinical features of each may be shortly stated, whilst the important observation made by Sangster (22) is considered more fully.

The patient seen by Fox (18) was a young man who "in childhood" suffered from annual attacks of cutaneous hyperæmia, followed by peeling of the entire skin in large flakes, as in scarlet fever. Recently he had similar attacks in a milder form, and during the ten days previous to exhibition the skin of almost the whole body had exfoliated, and the horny layer of the epidermis of the palms was absent in patches, leaving a thin reddened skin beneath. Shreds or flakes of epidermis were still adhering, and could be peeled off without pain.

Preston's patient was an Englishwoman, aged 67, living in

New Zealand. Her complaint was of "old standing"; but with this exception, she had been healthy since childhood. She had small-pox when two years old, and a large bronchocele developed at puberty. She had a large family, all healthy children. "Since childhood" she had suffered at intervals of a month or six weeks from the following occurrences: "After a day or two of slight feeling of *malaise*, the skin of every part of the body comes away in 'casts,' and the cuticle, which separates from the extremities, does so in one entire and sometimes unbroken piece, resembling a 'glove' or 'stocking.' The new skin beneath has the appearance of ordinary skin after desquamation, and 'lasts her,' to use her own expression, for several weeks, when it begins to get irritable and inflamed." Preston was able to watch her closely during one of these desquamating epochs, which, by the way, were only occasionally (and accidentally) associated with the menstrual flow, which was normal; and he noted that the exfoliated skin was of the thickness of ordinary cuticle, and was in places as tough as the lining of a hen's egg. The first attack the patient could remember was at the age of seven years.

The case reported by Frank and Sandford (20) was even more remarkable than the above. The patient, a man 33 years of age, was well developed, had never required medical attendance, had a normal skin without birth-marks, and had never had any of the eruptive fevers. He was the second of a family of thirteen, all of whom were alive and in good health. His father and mother were also living. He had had two slight attacks of gonorrhœa. During his mother's pregnancy nothing peculiar occurred, but his birth took place in the open woods at night during the Kansas and Missouri trouble. Seven months after his birth "he was taken suddenly ill, vomited, became hot and feverish, and in a few hours the entire surface of the body was scarlet-red." The symptoms increased for three or four hours, then subsided; but on the fourth or fifth day afterwards the entire cuticle was cast off, and a little later the nails of fingers and toes were also shed. This process was repeated every year on the same date (July 24th), *i.e.*, thirty-three times; and although he was seen by physicians from curiosity, he was

never treated for it. The writers then describe with full details one of these annual sheddings of the epidermis. The paroxysm begins abruptly, usually at 3 P.M., never later than 9 P.M. There is a feeling of lassitude, followed by muscular tremors, nausea, vomiting, and a rapid rise of temperature; and the skin and mucous membrane of the tongue and mouth become red and inflamed. The skin is hot and dry, no perspiration appearing till after the cuticle is exfoliated. The acute symptoms begin to subside in three or four hours, and disappear in twelve hours; and the redness of the skin passes away thirty-six hours later. In early life the shedding of the cuticle began on the second or third day, and ended on the fifth; but in recent years the process has occupied ten or twelve days. The epidermis is shed in large pieces, coming off the hands and feet in the form of "gloves" and "moccasins." The nails separate about a month after the acute stage. Microscopic examination of a piece of skin, removed under cocaine from the arm, showed nothing abnormal.

Sligh's patient (21) was the same man as Frank and Sandford described; but the former was present at the annual desquamation in the year after that in which the latter writers saw the shedding process. Some additional facts were noted by Sligh; the patient stated that he had missed peeling once when suffering from gonorrhœa; the skin did not peel off in an unbroken piece when the premonitory symptoms were prolonged, and under the epidermis small blebs filled with clear fluid appeared before exfoliation commenced. The new cuticle that formed was as soft and tender as that of a new-born babe; but it rapidly became hard, and the patient was fit for manual labour again.

Sangster's recently noted case (22) is a somewhat problematical one, which will doubtless be regarded by many as ichthyosis, although the absence of hyperkeratosis and papillary hypertrophy led the observer to doubt this and to call it "keratolysis exfoliativa." The patient, a man 24 years of age, was one of a family of ten, with no history of syphilis. His brothers and sisters showed no skin affection like that exhibited by the patient. When he was in the third week of life desqua-

mation was noticed, and at the end of the third year this had become generalized. The condition of the skin had persisted up to the present time; but three or four times every year he had "attacks," during which the condition was much aggravated and exfoliation much more active. There had never been bullæ on the skin, and perspiration occurred at all times on the palms and soles, and in hot weather over the whole body. The skin had everywhere a dusky tint, whilst in places it was of a brownish hue. Two varieties of surface appearances could be recognised. There were extensive tracts (*e.g.*, on the extensor aspects of the arms, outside of the thighs, etc.) of "harsh, cracking-like epidermis, much of it thickened and divided up into small quadrate areas of heaped-up sebaceous *dibris*. . . . Where not so thickened the skin felt brittle and paper-like, as if the epidermis were dead, and had lost organic connexion with the deeper layers below. On picking up the partly-detached margin of epidermis, the latter could be peeled up in sheets three to four inches square, without pain or much inconvenience to the patient. The surface beneath was then seen greyish-white, smooth, soft, and even, without any appearance of protruding papillæ; to the touch there was a slight stickiness. Within some hours the denuded surface became hyperæmic, and even of a brilliant red colour, and the patient experienced smarting of the part, but nothing of the nature of a crust formed, and the epidermis gradually lost its red appearance, became firm, and settled down into the same condition as that removed." In addition to the areas described above, there were others in which the surface presented "a broken-up, ragged appearance made up of—(*a*) tags of partly-detached epidermis; (*b*) flake-like, papery scales, with edges turned up; these flakes had a stuck-on appearance, and varied in shape and size, often somewhat angular, and nearly an inch in longest measurement; (*c*) intervening recently denuded skin, looking red and inflamed." There was no exfoliation on the palms and soles, where the epidermis was thickened, sodden, and bathed in sweat. The lesion was most marked on the back, abdomen, buttocks, and outer side of the thighs; but was also evident on even the scalp and glans penis. There was no alteration

of the hairs and nails. Sangster concluded that the condition was a malformation of the skin, and congenital; and that it was essentially non-inflammatory, having to do with faulty developmental changes in the upper layers of the epidermis. He thought that it was not ichthyosis; but it might be more closely allied to the milder types of that disease in which the papillary hypertrophy was not so marked.

Nature and Mode of Origin.—It is rather difficult to come to any conclusion with regard to the pathogenesis of the affection described; but it may be that Sangster's case at any rate is allied to the mild form of foetal ichthyosis discussed in Chapter VIII. With regard to the patients seen by Fox, Preston, Frank, and Sligh, it is evident that they were the subjects of a keratolytic process occurring periodically, and commencing so far back in infancy as to suggest an antenatal peculiarity of the skin, or at least of its epidermic layers. Sligh (21) regarded his case as probably one of dermatitis exfoliativa, but without the exhaustion supervening on recurring exfoliations or the changes noted in the skin in chronic cases of this disease. Crocker (*loc. cit.*) has placed this curious condition of the skin under the heading "Pityriasis," and seems to consider it as allied to the annual casting of the thickened epidermis in tylosis palmæ, an instance of which he has had under his charge. I have grouped it among the keratolyses in close proximity to desquamation in the living foetus and dermatitis exfoliativa neonatorum; but it must remain for some time a dermatological enigma and curiosity.

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CHAPTER XIII.

GENERAL CONSIDERATIONS.

CLASSIFICATION OF CONGENITAL CUTANEOUS AFFECTIONS : SCHEMES OF BAZIN, UNNA, AND AUTHOR ; REMARKS ON THE DIFFICULTY OF INVESTIGATING AND ARRANGING THESE DISEASES. SYMPTOMATOLOGY OF CONGENITAL SKIN DISEASES : GENERALIZATIONS. THE VALUE OF EMBRYOLOGY IN THE ELUCIDATION OF CONGENITAL SKIN DISEASE, AND *vice versa.*

IT will be convenient at this stage in the consideration of congenital or foetal skin diseases to make some general observations on the classification and characters of these maladies. I may, however, first advert to the method in which I am discussing antenatal maladies in order that an idea of the arrangement of the work may be obtained. It will be remembered that I divided all foetal diseases into four groups :* idiopathic, transmitted, traumatic, and toxicological. Now amongst the transmitted maladies, which fall to be discussed in a later volume, occur certain skin affections. These are the cutaneous manifestations of variola, scarlet fever, measles, typhoid fever, and erysipelas in the foetus, as well as the large group of the congenital syphilitic skin diseases. None of the above states have, therefore, been yet considered. Further, in the large group of idiopathic maladies I have reserved a place for congenital neoplasms, and amongst them must be placed the tumours of the skin and subcutaneous tissue, such as many of the nævi, congenital elephantiasis arabum (so-called), xanthoma cutis, congenital lipomata, myomata, etc. Of the idiopathic skin diseases left for consideration some have been already discussed, whilst others await notice in the next volume of this work.

I may here state what have been dealt with in this and in the first volume.

Diseases of the Subcutaneous Tissue—

General Foetal Dropsy or Anasarca, . Vol. I. page 102

* *Vide Diseases of the Fetus*, vol. i. p. 94.

General Cystic Elephantiasis of the Fœtus, Vol. I.	page	182
General Fœtal Obesity,	"	231
Sclerema (and Oedema) Neonatorum, .	Vol. II.	I
Subcutaneous Abscess in the Fœtus,	"	77
Atrophic State of the Subcutaneous Tissue,	"	79
Dermatolysis,	"	82

Diseases of the Skin—

Fœtal Ichthyosis (Grave Type),	"	88
" " (Mild Type),	"	130
Congenital Ichthyosis Hystrix (Major),	"	151
" " " (Minor),	"	157
Keratosis Pilaris,	"	169
Tylosis Palmæ et Plantæ,	"	176
Akrokeratoma Hereditarium,	"	183
Tyloma,	"	184
Clavus,	"	185
Cornu Cutaneum,	"	185
Fœtal Keratolysis,	"	188
Dermatitis Exfoliativa Neonatorum,	"	204
Periodical Keratolysis,	"	215

To put it briefly, I have considered the idiopathic diseases of the subcutaneous tissue, and such of those of the skin as have hyperkeratosis or keratolysis as their leading morbid change. There remain for discussion, therefore, congenital pemphigus (simplex); the congenital diseases of the hairs (hypertrichosis, paratrichosis, and hypotrichosis), of the sebaceous and sudoriparous glands, and of the nails; the pigmentary anomalies (albinism, nigrismus, and some of the nævi); and a few other skin affections, such as absence of the skin, cutaneous ulcers, xeroderma pigmentosum, congenital (?) eczema, and congenital (?) herpes zoster, which are probably due to antenatal morbid states. As I have already stated, the cutaneous neoplasms will appear later, as will also the transmitted skin affections.

It may, in the meantime, not be out of place to make here some general statements with regard to the whole group of the congenital idiopathic maladies of the subcutaneous tissue

and skin. Few dermatologists have considered congenital skins as a separate class, although all have discussed with more or less fulness the individual diseases. Of the few who have specially dealt with the group I may briefly refer to Bazin* and to Unna.†

In his article on the "Dermatoses," Bazin constantly separates for special notice a group of cutaneous maladies which he calls the "deformities." These he divides into two classes: acquired and congenital. The former class includes such cutaneous conditions as tatoo marks, and calls for no further reference here; the latter is almost, but not quite, synonymous with the idiopathic congenital skin diseases as discussed in this work. It is not necessary to distinguish sharply between diseases and deformities, for whilst some cases of congenital skin lesion are clearly diseases and others deformities, there are some which it would be exceedingly difficult to place in either one or other group exclusively. As Bazin himself says, most congenital skin diseases are cutaneous deformities; some are undoubtedly hereditary, whilst others, such as naevi, are caused by accidental conditions occurring after conception, but the distinction is not very important.

Looking at the dermatoses from the point of view of their pathological anatomy, it is evident that they may be divided into four classes which correspond to the four divisions of anatomy, *i.e.*,—descriptive, structural, regional, and embryonic. The cutaneous deformities correspond to the fourth division, to embryonic anatomy. Bazin (*loc. cit.*, p. 748) proposes further to arrange them in the following manner:—

Macular	Pigmentary	{ Hyperchromatoses Achromatoses Dyschromatoses	{ Pigmentary Nævi. Nigrismus. Lentigo. Albinism. Congenital vitiligo.
	Vascular		
	.		Nævus flammeus.

* Bazin, Article *Dermatoses* in *Dict. encyclop. des sciences méd.*, ser. i., vols. xxvii. p. 640, and xxviii. p. 1, 1882-3.

† Unna (P. G.), *Die Histopathologie der Hautkrankheiten*, pp. 1135-1202. Berlin, 1894.

Papular ("Bouttoncuses")	{ "Bouttoncuses"	{ "Nævus boutonneux," molluscum, warts.
Hypertrophic		
Exfoliative	Hypertrophic	Dermatolysis, cutaneous hypertrophy, elephantiasis arabum.
Ulcerative (very rare).		Ichthyosis. Ichthyosis pilaris.
Atrophic		Atrophy. Congenital absence of one or several layers of skin.
Cicatricial		Permanent cicatrices.

This scheme may be usefully compared with that proposed by Unna (*loc. cit.*, p. 1137). From the pathological standpoint he divides all skin diseases into six groups, the fifth of which, that of the malformations (Missbildungen), is the one with which we are concerned. It is as follows:—

A. Progressive Ernährungsstörungen.

I. Wucherungsgeschwülste.

- a. Syringadenom (Syringom).
- b. Akanthoma adenoides cysticum.
- c. Keratoma palmare et plantare hereditarium.
- d. Verruca dorsi manus et pedis.
- e. Nævi.
 - 1. Weiche Nævi.
 - 2. Harte und gemischte Nævi.
 - 3. Haarnævi.
 - 4. Riesennævi.
 - 5. Nævi lineares.
- f. Elephantiasis congenita fibrosa.
- g. Hypertrichosis congenita universalis.

II. Stauungsgeschwülste.

- a. Hyperkeratosis universalis congenita.
- b. Cysten.
 - 1. Dermoide.
 - 2. Atherome.
- c. Pigmentnævi.
- d. Nævi seborrhoici.
- e. Nævi angiomas, Elephantiasis congenita angiomas.
- f. Elephantiasis congenita lymphangiectatica.

B. Regressive Ernährungsstörungen.

- a. Congenitaler defekt der Haut.
- b. Albinismus partialis.
- c. Poliosis circumscripta.
- d. Atrichia localis, Hypotrichosis localis.
- e. Atrichia, Hypotrichosis congenita universalis.

I have not adopted either of the systems of classification which have been referred to, for the plan of this work did not fall in with them, and, indeed, Unna's scheme has only recently been published; but I have arranged the congenital skin diseases (including the "cutaneous deformities" of Bazin and the "Missbildungen" of Unna) in the following way. I omit the other diseases (those of the bones, viscera, etc.) in order to make more evident the cutaneous affections.

I. Idiopathic Diseases.

A. Of the Subcutaneous Tissue.

(Desmoses. Chorio-Blastoses.)

I. Cœdematous.

a. Général Fœtal Dropsy—Hydrops Fœtus universalis.

b. Elephantiasis congenita cystica universalis.

II. Hypertrophic.

a. Fœtal Obesity—Obesitas Fœtus universalis.

b. Sclerema (and Cœdema) Neonatorum.

III. Inflammatory. Subcutaneous Abscess in Fœtus.

IV. Atrophic.

Atrophy of Subcutaneous tissue.

Dermatolysis (?).

B. Of the Skin.

I. Epidermidoses.

1. Keratoses—Hyperkeratoses.

(1.) Ichthyosis Fœtalis { Grave Type.
Mild Type.

(2.) Ichthyosis Hystric congenita.

Major Degree.

Minor Degree—Nerve Nævus.

(3.) Keratosis Pilaris.

2. Acanthoses—Hyperacanthoses.

- (1.) Tylosis Palmæ et Plantæ. Keratoma palmarum et plantarum hereditarium (Unna).
- (2.) Akrokeratoma Hereditarium (Neuburger).
- (3.) Tyloma (congenital).
- (4.) Clavus (congenital).
- (5.) Cornu Cutaneum (congenital?).

3. Keratolyses.

- (1.) Keratolysis Fœtalis. Epidermic Desquamation in the living Fœtus.
- (2.) Keratolysis Neonatorum. Dermatitis exfoliativa neonatorum (?). (Possibly rather an angio-epidermidosis).
- (3.) Periodical Keratolysis.

4. Trichoses.

- (1.) Hypertrichosis congenita { universalis.
 localis (?).
- (2.) Hypotrichosis congenita { universalis.
 localis.
 (Atrichia universalis et localis.)
- (3.) Paratrichoses.
 - a. Trichorrhesis nodosa.
 (Monilethrix.)
 - b. Poliosis circumscripta — Canities congenita localis.

5. Chromatoses.

- (1.) Hyperchromatoses.
 - a. Melanoderma.
 - b. Nævus pigmentosus.
- (2.) Achromatoses.
 - a. Albinismus universalis.
 - b. , partialis (in negro).

6. Onychoses.

- (1.) Hyperonychia — Onychauxis (congenital).

(2.) Paronychoses.

Onychogryphosis congenita.

(3.) Hypo-onychoses—Congenital Absence or Atrophy.

II. Cryptoses.

1. Steatoses.

a. Hypersteatoses—Seborrhœa congenita.*b.* Asteatoses—Xeroderma congenita.

2. Idroses.

a. Hyperidrosis congenita.*b.* Paridroses—Hæmatidrosis congenita (?).*c.* Anidroses. Anidrosis congenita (?).

3. Crypto-stenoses.

a. Comedo.*b.* Milium.*c.* Acne (congenital).

4. Congenital Fibro-sebaceous Disease (Crocker).

III. Angio-epidermidoses.

1. Angiotic Acantholyses.

a. Pemphigus (congenital non-syphilitic).*b.* Pemphigus neonatorum.

2. Herpetic Dermatoses.

Fœtal Herpes zoster (Hutchinson).

3. Eczematous Dermatoses.

Congenital Eczema (Robinson).

4. Angiotic Parakeratoses.

Psoriasis (congenital ?).

IV. Neuritic Dermatoses.

1. Urticaria (congenital ?).

2. Urticaria pigmentosa (Sangster).

3. Hebra's Prurigo (Hutchinson).

4. Xeroderma pigmentosum (Kaposi).

V. Atrophic Dermatoses (Adesmoses).

1. Congenital Absence of Skin (H. v. Hebra).

2. Congenital Atrophy (*v. Ichthyosis fœtalis.*)
(*Atrophoderma striatum et maculatum.*)
3. Cutaneous Ulcers (congenital?).

VI. Hæmorrhagic Dermatoses.

1. Congenital Purpura.
2. (Hæmophilia).

II. Cutaneous and Subcutaneous Neoplasms. Desmoses.

1. Nævi (Hard, Soft, Hairy, etc.).
2. Elephantiasis congenita $\left\{ \begin{array}{l} (a) \text{ fibrosa.} \\ (b) \text{ angiomatosa.} \\ (c) \text{ lymphangiectodes.} \end{array} \right.$
3. Fibroma Cutis (congenital).
4. Lipoma " "
5. Xanthoma " "
6. Myoma Cutis (congenital).
7. Keloid (congenital, Bryant).
8. Adenoma Cutis (congenital).
9. Angioma Cutis (congenital), etc.

III. Transmitted Diseases.

A. Exanthematous Epidermidoses and Dermatoses.

1. Fœtal Variola.
2. Fœtal Scarlatina.
3. Fœtal Morbilli.
4. Fœtal Varicella.
5. Fœtal Typhus.
6. Fœtal Typhoid.

B. Phlegmonous Angiodesmoses.

1. Fœtal Erysipelas.
2. Fœtal Puerperal Fever (Sepsis).

C. Granulomatous Paradesmoses.

1. The Syphilodermata $\left\{ \begin{array}{l} (1) \text{ Macular, (2) Papular,} \\ (3) \text{ Squamous, (4) Vesicu-} \\ \text{lar, (5) Bullous, (6) Pus-} \\ \text{tular, (7) Tubercular.} \end{array} \right.$
2. Elephantiasis Græcorum congenita (?).

Such is the scheme of classification of congenital skin diseases which I venture to propose, and of which I am making use in this work. It is, as will be recognised, founded to a large extent upon the plan of arrangement of cutaneous affections in general which was proposed by Auspitz, and modified and elaborated by Bronson;* but I have departed from that system in several details in order to suit the requirements of my own classification of all antenatal maladies into idiopathic, transmitted, traumatic, etc., and for other reasons which I need not particularise. I could not adopt the classifications of Bazin and Unna, for these writers were dealing solely with cutaneous deformities (*Missbildungen*), whilst I wished to include all congenital skin affections. With regard to the occurrence before birth of several of the diseases which I have placed in the scheme of arrangement some doubt may exist; but I have included only those affections congenital examples of which some dermatologist of authority has reported. I have admitted also one or two cutaneous maladies which, although not developed for some time after birth, are yet believed by competent authorities to be essentially due to congenital faults in the structure of the skin. Such a disease is xeroderma pigmentosum of Kaposi.

It is more than likely that in process of time still further additions will have to be made to the list of congenital skin diseases. In the meantime dermatologists rarely have an opportunity of inspecting the cutaneous maladies that are present at birth, whilst the obstetricians and general practitioners who have this opportunity either fail to avail themselves of it, or through the want of special training in the recognition and classification of such morbid states do not do full justice to their material. It is not, therefore, wonderful that our knowledge of antenatal skin diseases is defective; there is even under the circumstances cause for congratulation that it is as extensive as it is.

It will be noticed that I have not placed any of the dermatomycoses or parasitic skin diseases in my list of congenital maladies, for, although Paulini long ago reported a case of

* Bronson (E. B.), *Journ. of Cutan. and Vener. Dis.*, ii. p. 206, 1884.

scabies in the fœtus, there is not sufficient evidence in support of their occurrence *in utero*, and, indeed, Paulini's case was probably not parasitic in its nature at all.

With regard to the symptomatology of congenital skin diseases Bazin (*loc. cit.*) makes some generalisations. Of course it must be borne in mind that he was only considering the smaller group of the cutaneous deformities. He points out that they are all stationary dermatoses, cutaneous affections arrested in process of evolution, and he arranges their characters under six headings. They (1) are local or universal in distribution ; (2) are not accompanied by itching (pruritus) ; (3) do not usually cause any derangement of the general functions of the system ; (4) do interfere very markedly often with the functions of the skin ; (5) have a slow progress ; and (6) have an indeterminate duration.

From the study of those idiopathic congenital skin diseases which have been already described it will be noted that the general characters ascribed to the whole group by Bazin are in many cases quite applicable ; but in one or two details occasional exceptions occur. Thus in the cases of grave foetal ichthyosis death within a few days followed practically constantly, although it is to a large extent true that the general functions of the patient were affected secondarily rather than primarily. Again, itching cannot be said to be invariably absent in all the cutaneous deformities ; but it is so frequently so, that the general statement to that effect may be allowed to stand. In other points the generalisations of Bazin may be accepted for the group of the deformities, always bearing in mind that they do not at all apply to the transmitted skin diseases, which are, indeed, symptoms rather than pathological entities. Further, the general characters enumerated by Bazin may be usefully but carefully employed to aid in the diagnosis of congenital from non-congenital skin diseases. It may be added that antenatal cutaneous maladies are often hereditary, but then this is true also of skin diseases that are rarely, if ever, present at birth. Further, the form of heredity exhibited is rather that of family prevalence, which is indeed often very marked, or of dissimilar than of direct and similar transmission of

any one malady from parents to children. This is a conclusion which may justly be drawn also with regard to deformities affecting other parts of the body than the skin, and very markedly with regard to teratological phenomena of a gross type.

As has been pointed out already by Bazin, much light may be expected to be thrown upon the mode of origin and nature of congenital cutaneous diseases by the study of the embryology of the skin; and conversely it may confidently be anticipated that the investigations of foetal morbid states of the skin will clear up problems in its development. This has been abundantly proved with regard to deformities of every other part of the body; for instance, the investigation of the evolution of the face has served to elucidate the mode of origin of hare-lip, macrostoma, branchial fistulæ, and the like; whilst conversely the study of these deformities has directed attention and guided research to many of the steps in the development of the various processes and fissures which are transitorily evident in the facial region during embryonic and foetal life. With regard to cutaneous disease in relation to the embryology of the skin, comparatively little has yet been accomplished, and a surely fertile field of investigation here lies ready to reward the dermatologist who is also an embryologist. Already some little progress has been made, and, as I have pointed out in speaking of general foetal dropsy, the peculiar characters of the dropsical infiltration (gelatinous rather than serous) in these cases is, I think, to be explained by the stage of development reached by the subcutaneous tissue at the time when it was attacked by this intra-uterine malady. Further, it has suggested itself to me that the investigation of the epitrichium in the human and other embryos may throw some light upon the mode of origin of foetal ichthyosis, whilst possibly the microscopical examination of the skin in that antenatal malady may give indications of value concerning the embryology of the epidermis and epitrichium. Similar suggestions might be thrown out regarding the mutual usefulness of research into the embryology of pigment formation in the skin, and into the histological and chemical characters of nævi.

Finally, the fact recently demonstrated by Porak,* that arsenic given to a pregnant animal is found stored up in the skin of the fœtus *in utero*, has an importance which can scarcely be over-estimated in its bearings upon dermatological therapeutics. But I must reserve further enquiries into this tempting sphere of investigation till I have completed the consideration of all the maladies of the skin which may justly be termed congenital or antenatal.

* Porak, *Archives de Méd. expér. et d'Anat. path.*, p. 192, March 1894. (Abstract in Ballantyne's *Teratologia*, i. p. 211, 1894.)

ADDENDA.

[*Note*.—Since the first volume of this work was published, and whilst the sheets of the present instalment have been in the press, some further contributions to the subjects dealt with have appeared: these, along with a few references which had been overlooked, are noted below.—J. W. B.]

HISTORICAL SKETCH OF THE DISEASES OF THE FœTUS.

Vol. I. p. 43, 5th line from top: for *performed* read *preformed*.

Vol. I. p. 64, 10th line from bottom: the dissertation "De Partu difficili ex Hydrope Fœtus" attributed to *J. C. Gehler* was written by *S. D. Naumannus*, *J. C. Gehler* being the President.

Vol. I. p. 79: *insert*, Corradi, in his historical work, "Dell' Ostreticia in Italia," published at Bologna in 1877, has summarised the contributions that Italian writers have made to such subjects as fœtal fevers, variola, vermes, maternal impressions, congenital enchondroma in cranium, fœtal hydrocephalus, ascites, struma, sacral tumours, etc.

GENERAL DROPSY OF THE FœTUS.

Vol. I. pp. 160–164. To the numerous references to cases of general fœtal dropsy given on pp. 160–164 several others must be added.

DAMMANN (P.)—*Ein Fall von Rhachischisis anterior et posterior mit Hernia diaphragmatica und universalem Hydrops der Frucht.* Inaug. Dissertation. Berlin, 1882. (This paper contains the details of a female fœtus with general dropsy and several other malformations; the dropsy was ascribed to pressure on the vena cava by the diaphragmatic hernia.)

GRIFFITH (W. S. A.)—“Dropsy of Pregnancy. Dropsy of the Fœtus in connexion with Dropsy of the Mother, and its

importance in the induction of Labour in such cases," *Brit. Med. Journ.*, vol. i. for 1889, p. 68, Jan. 12, 1889.

RIBEMONT-DESSAIGNES (A.)—"Contribution à l'étude de la macération chez le fœtus vivant," *Annales de Gynécologie*, xxxii. p. 8, 1889. (This article contains full details of five cases of general foetal dropsy. They are referred to also in the chapter on Foetal Keratolysis in the present volume.)

BALLANTYNE (J. W.)—"On Two further Cases of General Dropsy of the Fœtus," *Trans. Edin. Obstet. Soc.*, xviii. 215, 1892-93. (These two cases were the fourth and fifth specimens of this foetal malady examined by me.)

TAURIN (A.)—"Hydropsie généralisée d'un fœtus de six mois avec placenta extrêmement œdématisé," *Bulletins et Mémoires de la Soc. Obstét. et Gynécol. de Paris*, p. 10, 1893; also in *Archives de Tocologie et de Gynécologie*, xx. p. 153, 1893.

BALLANTYNE (J. W.)—"Two Cases of General Dropsy in the New-born Infant," *Archives of Pediatrics*, xi. p. 137, 1894. (My sixth and seventh specimens.)

GRIMSDALE (T. B.)—"General Foetal Dropsy," *Liverpool Medico-Chirurgical Journal*, p. 225, January 1895. (Maternal œdema and albuminuria; hydramnios; placenta large, soft, and friable.)

GENERAL CYSTIC ELEPHANTIASIS OF THE FŒTUS.

Vol. I. pp. 218-219. Add the following references:—

MARTIN.—"Kind mit mehrfachen Kysten, etc.," *Monatsschrift für Geburtskunde*, xx. p. 170, 1862.

FEHLING.—*Archiv für Gynäkologie*, x. p. 188, 1876.

MARTIN.—*Zeitschrift für Geb. und Gyn.*, i. p. 51, 1877.

BODE.—"Elephantiasis congenita mollis universalis," *Centralblatt für Gynäkologie*, xvi. p. 963, 1892.

CONDAMINE.—"Angeborene Cysten," *Centralblatt für Gynäkologie*, xvi. p. 912, 1892. An abstract from *Prov. méd.*, Jan. 9, 1892.

SCLEREMA NEONATORUM.

Vol. II. pp. 65-76. Add the following references:—

BENEDICKS (N.)—*De telæ cellulosæ indurazione in neonatis.*

Diss. Inaug., 1807.

SANNES (A.)—*De indurazione telæ cellulosæ infantum recens natorum.* Diss. Inaug. med. Groningæ, 1843.

MIRAS (J. M.)—*Le Sclérème des Nouveau-nés.* Thesis, Paris, 1894.

DUPARCQUE.—“Mémoire sur l’Endurcissement du Fœtus,” *Nouv. Bibliothèque médicale, Journ. de Méd. et de Chir. pratiques*, iii. p. 333. Paris, 1828.

This memoir, unfortunately overlooked by me when I was writing the chapter on Sclerema, is so important that it is necessary to give a summary of it here. Duparcque, after alluding to the observations of Chaussier, Andry, Baumes, Denis, Underwood, Hufeland, and others, goes on to point out that these authors are either silent concerning the occurrence of sclerema before birth, or else make very incomplete allusions to it. In order to fill up the gap thus left in our knowledge of the disease, he proceeds to relate the following case, which he believed to be similar to that reported by Usenbenzius in 1718.

The mother, 26 years of age and a ii.-para, felt foetal movements when four and a half months pregnant, but they were feeble and sluggish compared with those felt in her first pregnancy. At the ninth month of pregnancy, but still twenty days before the full term, the foetal movements ceased to be felt, and the mother became conscious of a firm inert mass in her abdomen which moved only with changes in the position of her own body. Eight days later (March 26th) labour pains began. The cervix was not dilated; and when Duparcque placed one hand on the mother’s abdomen, and the fingers of the other in the vagina upon the presenting foetal head, an unusual degree of resistance was experienced. Next day the membranes ruptured, and liquor amnii stained with meconium escaped. The head presented, but the uniform resistance of the scalp masked the sutures and fontanelles, so that the position could

not be recognised. In spite of powerful pains the head did not lose its spherical form, and no caput succedaneum formed, neither was there any folding of the scalp. After some time, and with great difficulty, the head was born, occiput to the front. The head was immovable on the neck ; so fixed was it that it was difficult to make it execute the movements of flexion and rotation. With considerable manual traction the shoulders were born ; but the large size of the abdomen and the fixed position of the legs further delayed delivery. The child, a male, was at length born. It was dead.

The umbilical cord was very thick (10 to 12 lines in diameter) ; it was 16 inches long, and was as firm as a piece of whalebone. It had a clear white colour and contained a large quantity of the gelatin of Wharton, which was more liquid than usual. The thickened arteries had a nearly cartilaginous firmness which ceased at the placenta on the one hand, and at the umbilicus on the other.

The infant measured 13 inches in length, and it was so rigid that one could, by grasping it by a hand or a foot, hold it out like a piece of wood. It had the appearance of a foetus of seven and a half months. Duparcque was not able to note whether its temperature was less than that of infants born dead, but the cooling of the body was rapid. The epidermis was macerated, and had separated over a large part of the body, and the skin was of a reddish-brown. The abdomen was enormously distended ; but the limbs had a dried-up appearance, and some force was needed to move them at the joints. The skin resisted pressure all over the body, and seemed as if fixed to the underlying parts.

Twelve hours after birth the autopsy was made. The body was quite cold, although it had been wrapped up in cloths. There was a little less general firmness, and the joints were a little more easily moved. On dividing the skin a little reddish serum exuded, and on cutting down to the bone near the knee that joint became much more mobile, so that the rigidity of the muscles was not the only cause of the immobility. The cellular tissue showed no fat ; the muscles were of a dark colour, and the veins were engorged with black blood. The abdomen con-

tained more than a pint of reddish serum. The stomach and intestines were contracted, and the latter contained little meconium. The mesenteric vessels were congested. The liver was a little larger than is usual at this age, had a dark red colour, and the gall bladder contained a small quantity of yellow fluid. There was a little reddish serum in the pericardium and in the pleural sacs ; the lungs were dark and firm ; the foramen ovale was nearly obliterated, and the ductus arteriosus normal ; whilst the heart, great vessels, and viscera were all much congested. The cranial bones were not united, and the fontanelles were still largely membranous ; there was a small quantity of sero-sanguinolent effusion in the dura mater, spinal canal, and ventricles. The cerebral sinuses and vessels were engorged with blood, and the brain substance was rather firm in consistence.

Duparcque pointed out the resemblance which his specimen bore to that described by Usenbenzius, noting, however, this difference, that in the latter case the infant lived for twenty-four hours, whilst in the former it had been dead for several days.

After further interesting notes on sclerema in general, and on the above case in particular, the author tabulated a series of conclusions which may here be stated.

1. The infant whilst still *in utero* is liable to be affected with sclerema ("endurcissement") and rigidity.
2. This condition affects foetuses of from three to five months, as well as infants nearer the full term.
3. The characteristics of the condition are—(a), uniform firmness and rigidity of the whole body ; (b), a dark colour of the skin ; (c), increase in the size of the trunk, whilst the limbs remain normal or are even diminished in bulk.
4. It is caused by an absolute or relative superabundance of blood engorging the circulatory apparatus and stagnating in the capillary system.
5. From this superabundance and stasis there result—(a), vascular engorgement of the tissues, especially those rich in capillaries, such as the liver, lungs, muscles, and skin ; and (b), the more or less abundant exhalation or transudation of a sanguinolent serum into the cellular and serous cavities.
6. The rigidity of the foetus and the firmness of its organs

and tissues depend upon (*a*) the mechanical distention of the tissues by the superabundance of blood, by serous infiltrations and exudations; (*b*), the retention of the coagulable part of the blood in the extremities of the capillary system; (*c*), the exercise of the contractility of the tissue not counterbalanced by vital expansibility; and (*d*), the tetanic contraction of the muscles.

7. The engorgement of the circulatory apparatus of the fœtus is due (*a*) sometimes to a plethoric state of the mother, causing "par contre-coup" plethora of the uterus, placenta, and infant; (*b*), uterine congestion of a menstrual type, or due to physical shocks or moral emotions; or (*c*), to an obstacle to the return of the blood from the fœtus to the placenta. In these circumstances the plethora is general and absolute. It is only local and relative when there is an obstacle to the return of the blood from the right to the left cavities of the heart.

8. The umbilical cord shows a hardening and rigidity similar to these states in the fœtus, and these are due to excess of Wharton's jelly, and possibly to the absence of the *vasa propria* of the cord.

9. From the nature of the causes of the sclerema of the fœtus and the mechanism of its development, this state must be regarded as pathological, and not as a cadaveric phenomenon.

10. The infant may be born before this state is so far advanced as completely to destroy life, but it cannot long survive its birth.

11. If death occur *in utero* (as is the usual result), and if expulsion does not immediately occur, then the cadaver may retain its firmness and rigidity so long as it is enclosed in the membranes and protected by the liquor amnii. Decomposition alone causes the disappearance of the cadaveric rigidity.

12. Thus the explanations of the cause of sclerema usually given (*i.e.*, cold, localised diseases, etc.) are controverted.

13. The signs by which this state may be recognised while the fœtus is still *in utero* are—(*a*), More or less sudden cessation of fœtal movements; (*b*), the sensation of a firm mass in the body of the mother, moving with her movements; and (*c*), the discovery, by vaginal and abdominal palpation, of a firm and immobile body *in utero*.

14. This hardness, by masking the projections and depressions

of the foetal body, renders the diagnosis of the presentation and position difficult.

15. It gives negative or uncertain signs concerning the death or life of the foetus.

16. It causes delay and difficulty in labour by interfering with the normal mechanism.

Duparcque's case is a singularly interesting one, whilst his conclusions, although not all capable of withstanding criticism in the light of modern research in physiology and pathology, are yet very valuable. That the rigidity was not simply or solely foetal rigor mortis is, I think, proven by the state of maceration of the cuticle; whilst at the same time it is very striking that the author seems to have had in his mind the possibility of the existence of ante-natal cadaveric rigidity. Of course, as I have elsewhere pointed out, it is always difficult to be sure of the nature of any morbid change when it is accompanied by the signs of foetal death, yet I am inclined to believe with Duparcque that this was truly a case of ante-natal sclerema comparable to the observation made by Usenbenzius.

FœTAL ICHTHYOSIS (MILD TYPE).

Vol. II. p. 130.

GROSZ (GYULA) and TÖRÖK (LAJOS)—“Exfoliatio lamellosa neonatorum (Ichthyosis sebacea),” *Gyógyászat*, vol. for 1894, p. 423, September 9, 1894; also in *Annales de Derm. et de Syph.*, 3rd ser., vi. p. 104, 1895.

Since the part of this work dealing with Fœtal Ichthyosis was put in print, the above important communication by Grosz and Török has appeared, and a summary of it may here be given.

The mother, a i.-para, was healthy, gave birth on 23rd January 1894 to a well-developed infant, weighing 3850 grammes, and having a length of 56 cms. Its temperature after birth was 36°·4 C.

The authors first saw the infant on the following morning; and they then noted that the cranial circumference was 38 cms., that there was oedema of the occipital region, and that there were some excoriations on the scalp, caused by the forceps which had been required to accelerate labour.

The skin of the new-born infant was everywhere dry, shining, and of a clear brownish-yellow colour, specially marked on the anterior surface of the trunk, on the face, and on the limbs. The skin, which seemed as if painted with collodion, was traversed by quite superficial fissures in the regions of the abdomen, thorax, and extremities. The shining stratum separates at the margin of these fissures, and there is thus produced a fine lamellar border some centimetres in breadth, with its free edge towards the fissures. The dry and shining layer separates only at the level of the fissures ; everywhere else it adheres closely to the subjacent layer of the skin, although here and there it forms some fixed folds. At the level of the fissures, which have a width of from some millimetres to three or four centimetres, the skin deprived of its collodion-like covering can be seen ; it appears healthy, flexible, and of normal colour. It is impossible to produce desquamation by scraping. On the back the dry and shining stratum is not absent, but here the colour of the skin is rather red, and there are no fissures such as exist on the anterior part of the trunk. The hairy scalp is normal, and the hairs are well developed. The skin of the palms and soles is like that of the abdomen. There are no other anomalies (*e.g.*, deep rhagades, contractures, etc.). The stump of the umbilical cord has a fawn, brownish-yellow colour.

On January 24, the temperature was $36^{\circ}4$ C. in the morning, and $37^{\circ}3$ C. in the evening ; the infant is well, but has not taken the breast.

On January 25, the morning temperature was $37^{\circ}5$ and the evening $37^{\circ}8$. The infant sucks quite well. Lamellar desquamation has begun on the trunk.

On January 26, the morning temperature was $38^{\circ}1$. The infant weighed 3450 grammes. The cord had mummified. Since the night the child no longer sucked, and there was dyspnœa. The left eye was closed, the right half open. The right pupil was small, the left was large, and reacted only very slowly to light. The mouth was a little drawn to the left side, and there were occasional spasms of the limbs. The dyspnœa increased, and the infant died at 11 P.M.

At the autopsy (by Dr Minich), the skin was found in the

state above described. There was a hæmatoma in the right temporal muscle. There was haemorrhage at the level of the parietal tuberosities between the cranial bone and the periosteum. There was also haemorrhage between the dura mater and the subjacent tissues on the left side of the cerebrum and cerebellum. There was nothing abnormal in the internal organs.

The authors point out that their case shows the greatest similarity to a part of the description of "ichthyosis sebacea" given by Hebra, and later by Kaposi; but differs from it in not showing painful cracks of the mouth, rigidity of the mouth and nose preventing sucking, and consequent inanition leading to death. With regard to "fœtal ichthyosis" as described by Kyber, Hans Hebra, and others, it has also resemblances.

Grosz and Török believe that the two anomalies, so-called ichthyosis sebacea and ichthyosis fœtalis, are of an essentially different nature; and they give the following explanation of the former.

In new-born infants there is rather frequently noticed a pityriasisiform, furfuraceous, mealy desquamation. It occurs over more or less extensive areas. This is the natural desquamation of the new-born (Rayer), and it has no connexion with seborrhœa. In the treatise of Hebra and Kaposi, the description of an analogous state of the skin of the new-born is contained in the chapter on Seborrhœa, and it is said that the mealy and lamellar squames are formed by the drying of the vernix caseosa which has been imperfectly removed after birth. A short histological examination shows the falseness of this supposition, for the pityriasis-like squames are solely made up of horny cells, and contain sebaceous (fat) cells only at the level of the mouths of the sebaceous glands. What has taken place in the new-born, therefore, is simply a separation of the cells of the stratum corneum of the epidermis, a process which has been studied and explained by Kölliker (*Handbuch der Gewebelehre des Menschen*, vol. i., 1889), and which occurs several times *in utero*, and may persist for some time in extra-uterine life.

It is alongside of this "natural desquamation" of the skin of the new-born that the authors place "ichthyosis sebacea"; and according to their view it is erroneous to group it with sebor-

rhœa, and to explain it, as Kaposi does, as the result of a persistence after birth of the normal seborrhœa of the fœtus.

The authors believe that in their case the shining superficial layer was composed of the same elements as in the cases of desquamation of the new-born, and they also think that the condition would have entirely disappeared had not the infant unfortunately died of meningeal hæmorrhage due to the use of the forceps.

Their case, and along with it the cases of "ichthyosis sebacea," were nothing else than rare and unusual examples of the "natural desquamation" of new-born infants. This *physiological* desquamation of the embryo and new-born takes place usually in the form of a furfuraceous or finely lamellar pityriasis. It is rare to see so extensive and so complete a desquamation of the stratum corneum as in the cases of so-called ichthyosis sebacea, and in that of the authors; but one has several times noted a similar desquamation in analogous circumstances in the lower animals. Welcker has observed it in the embryo of the sloth, which is enveloped by the separated stratum corneum as by a second amnion. Under this layer the developing hairs project from their follicles, and on this account Welcker called it the epitrichium. The epitrichium is nothing but the stratum corneum of an early period of embryonic life, and it has been observed also in other animals. The desquamation of the embryo and new-born of the human subject represents the same process, differing only in the fact that the cohesion of the horny cells is less, and that, therefore, the horny layer breaks up. For cases such as theirs, the authors presuppose a more marked union of the horny cells, which leads to the formation of a horny envelope quite analogous to the epitrichium of certain animals.

In conclusion, Grosz and Török consider the state as a passing one, and not pathological in nature; it represents a variety of the physiological desquamation of the new-born. It is quite different from fœtal ichthyosis, which is an atrophic error of development; it cannot be compared with ichthyosis vulgaris, which is stationary; and it has no relation to seborrhœa. They regard the name ichthyosis sebacea as quite erroneous, and propose that of *exfoliatio lamellosa neonatorum*.

The above contribution is a very important one, and had not been published when I wrote my chapter on Ichthyosis Foetalis of the mild type. I agree with the authors in thinking that cases such as theirs ought to be separated from the grave type of foetal ichthyosis, and that they are not of the nature of seborrhœa; but I differ from them in certain particulars. I regard such cases as allied to foetal ichthyosis of the grave type, differing from it in degree rather than in kind. Ichthyosis, as is well known, is stationary, but so were many of the cases of the mild variety of foetal ichthyosis (*v. Chapter viii., p. 147*), and practically all the instances of the grave type.

There is one important conclusion that I am inclined to draw, and that is the possible connexion which may be regarded as existing between what I have called foetal and neonatal keratolysis and the cases of mild foetal ichthyosis. It is indeed probable that connecting links between these two groups of congenital skin disease may exist and may be capable of future demonstration.

With regard to the suggestion of Grosz and Török that the horny layer in foetal ichthyosis is comparable to the epitrichium of some of the lower animals, I may state that a somewhat similar idea had struck me (*v. p. 233*), an idea which I also expressed some time ago in a letter to Dr Ohmann Dumesnil of St Louis, who has kindly promised me a paper on the epitrichium for my *Quarterly Journal of Ante-natal Pathology*. I am, however, inclined to regard the horny layer of both the varieties of foetal ichthyosis as a persistent epitrichium.

TYLOSIS PALMÆ ET PLANTÆ.

Vol. II. p. 186. Add the following reference:—

SHERWELL.—“A Case of Congenital Idiopathic Tylosis,” *Brooklyn Med. Journ.*, p. 698, Nov. 1894. (A girl, 17 years of age, suffering from an extensive condition of tylosis involving the palms of the hands and plantar surface of the feet, which had existed to a slight extent since birth, but had become more noticeable since she was seven years of age; the condition was improving under the use of a salicylic and resorcin ointment.)

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